ST. VINCENT CHARITY MEDICAL CENTER

JOURNAL

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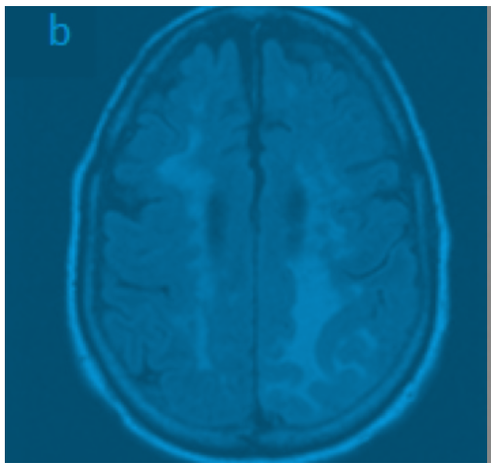

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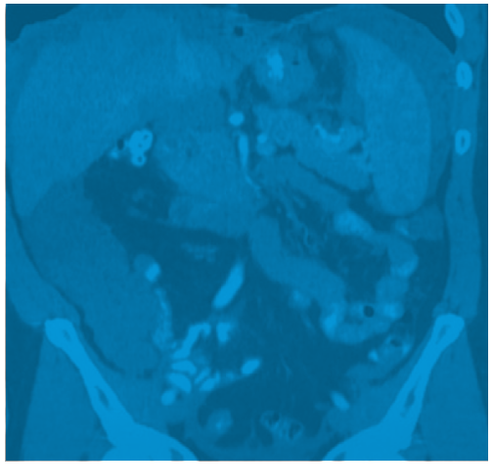

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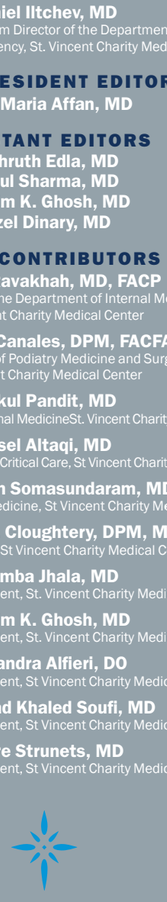

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MEDICAL CENTER

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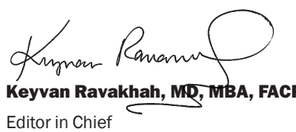
A MESSAGE FROM THE EDITOR

The 100th anniversary of the American College of Physicians (ACP) was recently celebrated in Boston, Massachusetts. The event hosted more than 6,500 internal medicine specialists and subspecialists. It included exciting, up-to-date and practical medicine lectures and hands-on exercises that impressed many of us while many outstanding experts talked of good medicine to hungry students and physicians.

However, if you ask me what the hallmark of the event was, my answer is "business." The most fascinating part of the event was the exhibition hall, a giant several thousand-square-foot arena filled with sponsors who paid thousands of dollars to our beloved American College of Physicians to have a chance in marketing their products, great or not, to us. There were plenty of buyers as well.

Medical students, residents, fellows and practitioners fought over shiny pens, pins and other cheap throw-away souvenirs while their brains were washed with the data on the best asthma, diabetes or hypertension medications in the market. Now the question is why this wholesome organization, the signature of integrity surrenders to industry as such. You may answer money and I agree with you partially.

I should mention that every attendee paid at least $500 fee for this three-day event. (I paid more than $700 myself.) I don't think American College of Physicians is going to declare bankruptcy soon but just coming back to the question I asked earlier, I think the better answer is greed. When you have enough but you want more, this is greed.



Keyvan Ravakhah, MD, MBA, FACP
Editor in Chief

We want to hear from you. Send your feedback to researchjournal@stvincentcharity.com

::: CLINICAL VIGNETTE :::

Anti-synthetase syndrome associated with cryptogenic organizing pneumonia

By **Nilamba A Jhala, MD; Kusuma Kurmayagari, MD; and Keyvan Ravakhah, MD**

INTRODUCTION

Anti-synthetase syndrome (ASS) is a heterogeneous autoimmune connective tissue disorder presenting as an inflammatory myopathy. ASS is usually associated with particular myositis-specific autoantibodies, which are specifically directed against the aminoacyl-t-RNA synthetase enzymes. The organs involved are numerous and diverse, and can vary from patient to patient. We present a case of ASS associated interstitial lung disease (ILD) and will discuss the clinical implications.

CASE DESCRIPTION

A relatively healthy 45-year-old African-American man presented with six-week history of progressively worsening dyspnea, non-productive cough, low-grade fever and fatigue. He also had associated pain and swelling of his extremities for about three weeks. In the meantime, he was evaluated once in outpatient clinic and twice in different emergency rooms (ER). He was treated as a case of community-acquired pneumonia (CAP) without any significant clinical improvement. Eventually he was admitted for further management of outpatient treatment failure CAP after his third ER visit.

On admission, he was afebrile, hypoxic (Pulse Ox 91% on room air) with sinus tachycardia of 117/min and BP 156/88 mmHg . Physical examination revealed dry crackles throughout bilateral lungs, proximal muscular weakness involving bilateral upper and lower extremities, mechanic's hands (Figure 1) and synovitis of MCP, PIP and DIP joints associated with trace pitting edema of all extremities.

There was no skin rash, mouth ulcers or palpable mass. The remainder of physical examination and review of system was unremarkable. Initial laboratory evaluation was remarkable for WBC of 12.5, CK > 26,000 K, ESR 72, myoglobinuria with positive rheumatoid factor and anti- Jo antibodies.

Chest radiograph revealed diffuse interstitial thickening in both lung fields, predominantly involving the lower lobes with associated small to moderate bilateral pleural effusions (Figure 2 & 3). HRCT revealed multifocal nodular ground glass opacities and confluent areas of consolidation with mild septal wall thickening and small to moderate bilateral effusions suggestive of acute ILD in a pattern observed as cryptogenic organizing pneumonia (COP) (Figure 4).

A diagnosis of ASS with COP was made and the patient was started on high-dose corticosteroids. Muscle biopsy was performed and was suggestive of polymyositis. Patient did not show much improvement with steroids, hence cyclophosphamide was added to his treatment regime. He slowly showed significant clinical improvement and was discharged | continued on p.17

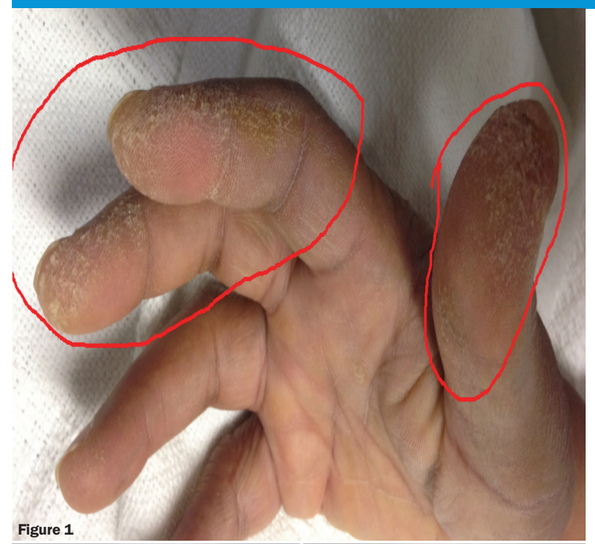


Figure 1

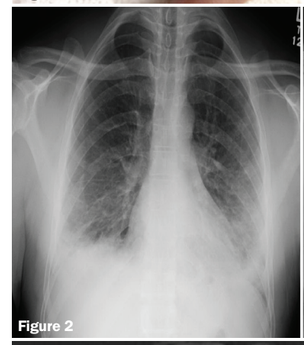


Figure 2

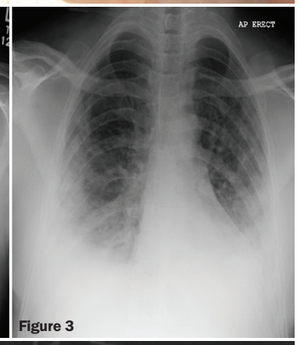


Figure 3

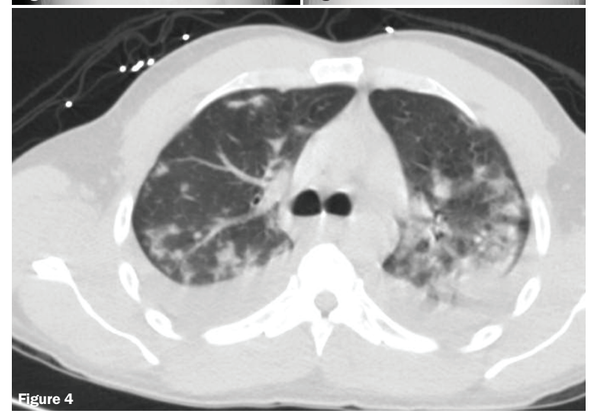


Figure 4

Figure 1: Mechanic's Hands
Figure 2 & 3: Initial CXR on 1st & 3rd ER visit showing worsening of bilateral patchy pulmonary interstitial infiltrates and effusions with diffuse interstitial thickening
Figure 4: HRCT showing bilateral multifocal nodular ground glass patchy airspace opacities and confluent areas of consolidation with a peripheral predominance and mild septal wall thickening with small bilateral pleural effusions suggestive of cryptogenic organizing pneumonia

Isolated Subtalar Joint Arthrodesis for Rearfoot Deformities:

A Case Series of Results from a Single Surgeon

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Corresponding: **Coleman O. Clougherty, DPM, MA PGY3, Mark C. Razzante, DPM, MA PGY3, Matthew M. Reiner, DPM PGY2, Grace Chuang, DPM PGY1**

ABSTRACT

Subtalar joint arthrodesis is commonly utilized for the correction of a variety of rearfoot deformities. The authors examined the postoperative patient satisfaction and functionality scores as well as radiographic analysis of surrounding joints on patients with an isolated subtalar joint arthrodesis. This procedure was performed by the senior (MBC) author on nineteen limbs (eighteen patients) utilizing a consistent operative technique and post-operative course at one institution over the course of seven years between 2007 and 2014. The AOFAS score improved from 47.68 (ffi 7.75) to 78.16 (ffi 9.07). SF-36 scores were above 73 for all categories with highest scores the in social and emotional role functioning categories of 91.45 and 98.25, respectively.

Comparing patient outcomes to radiographs, patients with preoperative osteoarthritis of adjacent joints had experienced an increase in arthritic pain postoperatively whereas those without preoperative osteoarthritis did not experience an onset of adjacent joint osteoarthritis postoperatively. The subtalar joint arthrodesis has proven to be a reliable procedure for a variety of rearfoot driven pathology in patients without concurrent adjacent joint osteoarthritis, however those with osteoarthritis in adjacent joints preoperatively should be informed of the potential of increased arthritic symptoms to adjacent joints postoperatively.

LEVEL OF EVIDENCE — *Level III*

INTRODUCTION

Isolated subtalar joint arthrodesis has been described as a surgical treatment option for a wide range of conditions affecting the lower extremities. Conditions include: calcaneal fractures, subtalar arthroses due to primary or post-traumatic arthritis, talocalcaneal coalitions, subtalar instability, adult acquired flat foot secondary to posterior tibial tendon dysfunction, talar fractures, subtalar joint dislocation, residual congenital deformity, inflammatory arthropathies[7], pes cavus, Charcot Marie Tooth, poliomyelitis, cerebral palsy, Reiter's syndrome, psoriatic arthritis, juvenile rheumatoid arthritis, rheumatoid arthritis, myelodysplasia, vertical talus, bone tumor, talipes equinovarus, avascular necrosis of the talus, talar cyst, and spina bifida[15]. Various techniques have been described including various screw orientations, variations forms of fixation, joint preparation, and graft augmentation.

The authors have retrospectively reviewed patients who have undergone isolated subtalar joint arthrodesis, to determine both clinical and radiographic outcomes after a mid-term follow up of 2-7 years by obtaining retrospective AOFAS scores pre and post operatively, SF-36 post-operative scores, and Kellgren-Lawrence Arthritis Grading Scales results. The patients included in this review were treated by a single surgeon (MBC) in one institution (SVCMC) with

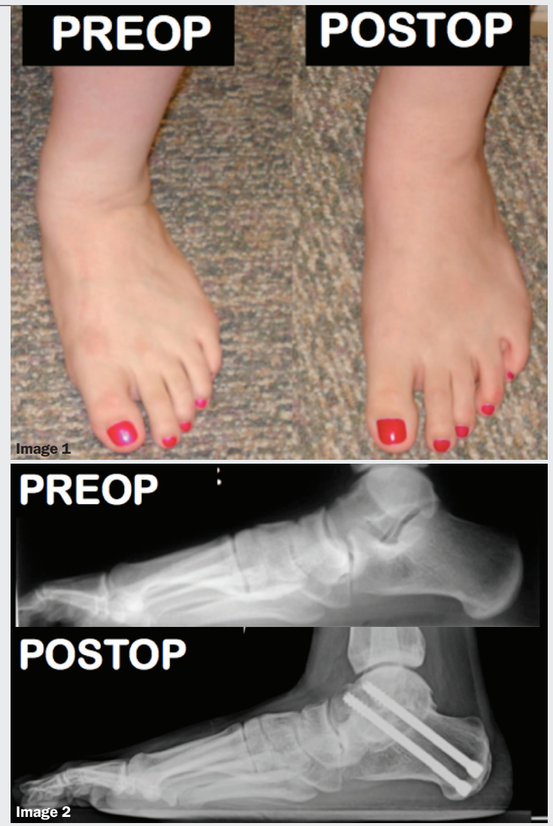
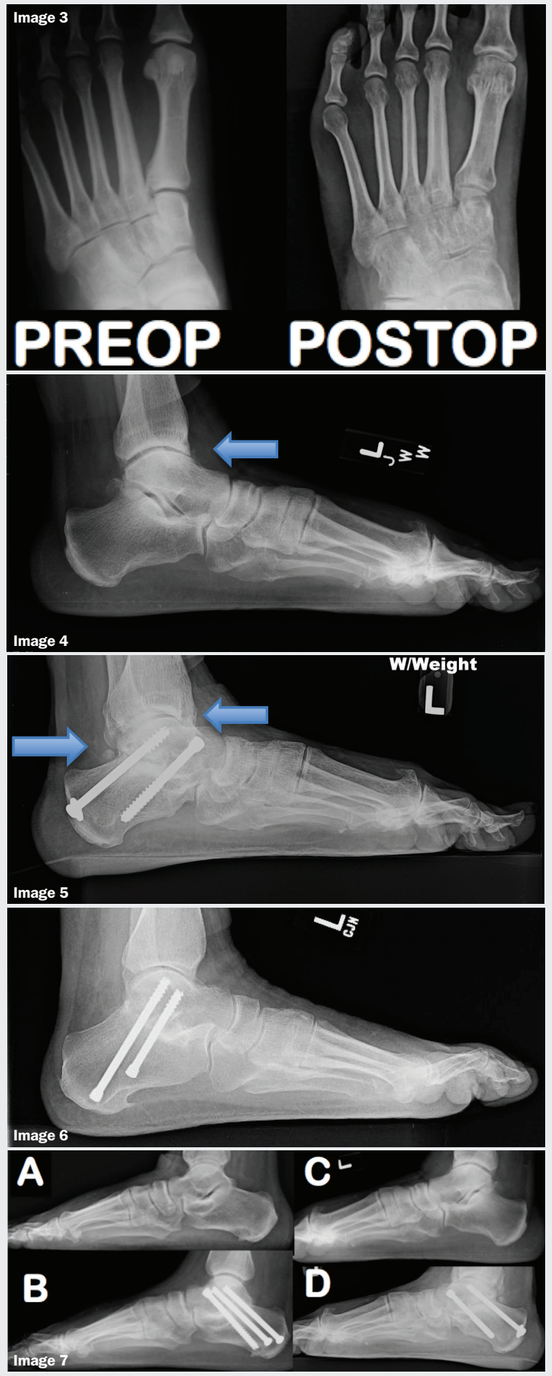


Image 1: Preoperative and Postoperative Clinical Weight Bearing Photo of Patient #7 showing correction in all three cardinal planes
Image 2: Preoperative and Postoperative Radiographic Weight Bearing Lateral Views of Patient #7
Image 3: Preoperative and Postoperative Radiographic Weight Bearing Anterior-Posterior Views of Patient #7
Image 4: Preoperative Radiographic Weight Bearing Lateral View of Patient #11 showing Preexisting Arthritic findings (arrow)
Image 5: Postoperative Radiographic Weight Bearing Lateral View of Patient #11 showing Exacerbation of Arthritic findings (arrows)
Image 6: Postoperative Radiographic Weight Bearing Lateral showing complete fusion 11 months after procedure of Patient #14
Image 7: Preoperative(A) and Postoperative(B) radiographs of Patient 1 without adjacent joint arthritis; Preoperative(C) and Postoperative(D) radiographs of Patient 2 showing advancement of pre-existing arthritis of adjacent joints
Image 8: Standard incisional approach with clear visualization of the subtalar joint's middle facet



the use of a single surgical approach with a similar internal fixation. All patients were followed postoperatively by the senior author.

MATERIALS AND METHODS

Following a comprehensive clinical and radiographic evaluation, a subtalar joint arthrodesis was deemed an appropriate treatment for the patients. When necessary, soft tissue procedures were included in the surgical plan, (recession or slide of the gastrocnemius aponeurosis, tendoachilles lengthening (TAL) or a posterior tibialis tendon repair). Two or three of the 6.5 mm cannulated screws were placed parallel to one another utilizing a percutaneous approach through the posterior plantar calcaneal heel pad.

Serial casting was performed when necessary for a total of six weeks of a non-weight bearing course on the operative limb. At that time, the patient was transitioned into a pneumatic fracture walking boot for an additional four weeks and permitted to weight bear in the device. At approximately 10 weeks post-operation, patients began ambulating in athletic shoe gear or in conjunction with a short articulating ankle-foot orthosis when appropriate for up to one year post-operatively.

Following Institutional Review Board approval, the retrospective chart review identified a total of 32 patients who underwent an isolated subtalar joint arthrodesis. After all eligible patients were contacted, a total of 18 patients (19 feet) were enrolled in the study. The remaining candidates were either unwilling or unable to participate due to location (7 patients) or unable to be reached (seven patients). The mean age was 45 years with a range of 21-64. There were 14 females and five males. Procedures were predominately performed on left side (13 limbs) with six right-sided operations. A total 18 ancillary soft tissue procedures were performed including the following: 12 gastrocemius recessions, one tendoachilles lengthening, four posterior tibial tendon repairs, and one peroneal tenotomy.

Patients were asked to be clinically evaluated in the office setting by the senior author and one of the corresponding authors, followed by the completion of a retrospective AOFAS score, a current AOFAS score, and an SF-36 questionnaire. Patients had their midfoot and ankle joints examined for residual or new arthritic findings followed by a gait evaluation. The patients then had radiographs performed of their operative limb in angle and base of gait. These images were compared to immediate postoperative films and graded utilizing the Kellgren-Lawrence Arthritis Grading Scale (Figure 1). Results were organized and tabulated (Tables 2-4).

RESULTS

Each of the 18 patients were examined by both the senior author and a corresponding authors. The patients were asked to complete AOFAS questionnaires for their postoperative condition, which were compared to preoperative scores tabulated based off chart

Figure 1: Kellgren-Lawrence Arthritis Grading Scale*

Grade	Characteristics
I	Doubtful joint space narrowing, possible osteophytic lipping
II	Definite osteophytes, definite joint space narrowing
III	Moderate multiple osteophytes, definite joint space narrowing, some sclerosis and possible deformity of bone contour
IV	Large osteophytes, marked joint space narrowing, severe sclerosis and deformity of bone contour
Standard Deviation	**Adapted from: Kellegren JH, Lawrence KS. Radiological assessment of osteo-arthritis. Ann Rheum Dis. 1957; 16(4): 494-502. Copyright 1957 BMJ Publishing Group Ltd. And the European League Against Rheumatism.*

review and patient input. The average preoperative AOFAS score was 47.68 with a standard deviation of 7.75, with scores ranging from 31 to 62. All patients had difficulty with ambulation and required an ankle-foot orthosis as a component of the non-surgical preoperative management by the senior author. The average postoperative AOFAS score was 78.16 with a standard deviation of 9.07, with scores ranging from 62 to 92.

All patients demonstrated an increase in AOFAS scores with improvement in pain, ability to increase their activity levels, and quality of life. The average increase in AOFAS scores was 30.47 with a standard deviation of 11.58, with scores ranging from 9 to 50.

The SF-36 scores were tabulated as seen in Table 4. The average Physical Functioning score was 77.37, Physical Role Functioning score was 75.26, the Bodily Pain score was 69.32, the General Health Perception score 81.32, the average Vitality score was 74.74, and the average Social Role Functioning score was 91.45. The average Emotional Role Functioning score was 98.25 and the average Mental Health score was 85.47. These results can be compared with the average SF-36 scores of the general population, which are displayed in Figure 2.

Each patient had preoperative, immediate postoperative, and mid-term follow-up radiographs taken of their surgical limbs. All radiographs were examined and evaluated for signs of arthritic findings such as joint space narrowing, peri-articular lipping or cyst formation, osteophytes, or adjacent joint sclerosis formation of the midfoot and ankle joints. The radiographs were analyzed using the Kellgren-Lawrence Arthritis Grading Scale (Table 3). Patients with preoperative arthritic findings had an increased propensity for progression of arthritic changes after the isolated subtalar joint arthrodesis. In patients who did not have preoperative adjacent joint arthritic findings, there was no evidence of advancement of arthritic changes, postoperatively. Consequently, no new adjacent joint arthritic findings developed during mid-term follow up in this cohort.

DISCUSSION

Many surgeons opt for joint preservation techniques in order to minimize adjacent joint arthritis, retain hindfoot motion, and avoid the possibility of revisional procedures secondary to nonunion of an arthrodesis. Astion and Deland demonstrated a mean 26% of the motion of the talonavicular joint, 56% of the motion of the calcaneocuboid joint, and 46% of the excursion of the posterior tibial tendon was maintained after simulated fusion of the subtalar joint. Heus et al examined the influence of subtalar and triple arthrodesis on the tibiotalar joint in 48 patients with a mean follow up of ten years. They utilized the Van Dijk radiographic grade scale which consists of the following: no abnormalities in Grade 0, evidence of cartilage damage in Grade 1, cysts and bone collapse in Grade 2, and joint space disappearance in Grade 3. There were no changes in 36 feet, and only 1 grade increase in 14 feet. Feet showing radiographic progression of arthritis had similar degenerative changes to the contralateral limb [5]. While there was high patient satisfaction in a case series of isolated subtalar fusions, there was a 36% and 41% incidence of mild radiographic progression of arthrosis in the ankle and transverse tarsal joints respectively[6]. Conversely, there was only an advancement of degenerative changes in adjacent joints in 14% after an average of 51 months [7].

While most evidence supporting isolated STJ arthrodesis in flatfoot deformity are case series or retrospective reviews, literature is scant with high level evidence for use of this procedure. Joint preservation versus joint fusion will always be a controversial topic as noted by Hiller et al[2]. Surgical planning is often influenced by individual training and is selected by surgeon preference. Correction of this subtalar joint malposition or peritalar subluxation is critical in restoring form and function of the foot, which consequently decreases pain. An important finding by Mann et al was that patients who underwent STJ arthrodesis for posterior tibial tendon dysfunction had a significant improvement in foot architecture with positive clinical outcomes, with 11 out of 44 patients who underwent this procedure for posterior tibial tendon dysfunction. There was a 93% satisfaction rate with an average AOFAS score of 89. The average follow up was five years, and a union rate of 100%[6].

Although a union rate was not reported, Yu et al offered insight as to execution and correction offered by subtalar arthrodesis. In 32 of the 48 of their subtalar arthrodeses, the following ancillary procedures: repair of tibialis posterior tendon, resection of tarsal coalition, peroneus brevis tendon lengthening, gastrocnemius recession, tendoachilles lengthening, modified Kidner procedure, ankle arthroplasty or synovectomy, and correction of painful digital deformities. With radiographic parameters, they found multiplanar correction when evaluating cuboid abduction, talar declination, talar uncoverage, and talar 1st metatarsal angle. They concluded that subtalar arthrodesis can aide in triplanar correction. Surgeons should expect to find minimal change in the calcaneal inclination angle according to their results and the authors recommend custom orthoses to accommodate any residual varus deformity or medial column stability[15].

Russotti et al noted a high satisfaction rate, as well as no radiograph findings of secondary degenerative changes in the hindfoot. Patients who were dissatisfied noted difficulty adjusting to uneven terrain. With their prospective trial, Fellmen et al further discredited the notion of adjacent arthritis. In fact, their results show that Chopart's joint and ankle joint range of motion was the same or better in 69% and 76% of cases respectively. The study also found a decrease in the consumption of pain medication as well as an increase in walking speed[20].

Johnson et al embraced the controversy of stage II posterior tibial tendon dysfunction with a detailed literature review of joint preservation osteotomies and joint fusion for this pathology along with a case series of 17 patients. Their procedure consisted of subtalar arthrodesis combined with spring ligament repair or reefing and flexor digitorum longus transfer to the navicular. Tendoachilles lengthening was also performed in a majority of the cases. There was 100% radiographic union occurring at a mean 10.1 weeks. There was improvement in talonavicular coverage, lateral talocalcaneal angle, and medial cuneiform height. Postoperative mean AOFAS scores were 82, and the mean Maryland Foot Score was 86. They stated that subtalar arthodesis allows for the ability to correct the rotatory subluxation of the calcaneus in relation to the talus[17].

Figure 2: Normal for the General United States Population, Total Sample*

Total Sample N = 6742	PF	RP	BP	GH	VT	SF	RE	MH
Mean	50.00	50.00	50.00	50.00	50.00	50.00	50.00	50.00
25th Percentile	46.51	47.06	41.83	44.83	45.85	45.94	48.10	44.38
50th Percentile	54.93	54.50	51.13	51.98	52.09	56.85	55.88	52.82
75th Percentile (median)	57.03	56.85	55.36	57.70	58.33	56.85	55.88	58.46
Standard Deviation	10.00	10.00	10.00	10.00	10.00	10.00	10.00	10.00
Range	14-58	17-57	19-63	16-64	20-71	13-57	9-56	7-65

*According to the U.S. Census 1998

Table 2: Increase in AOFAS Scores

PATIENT	CHANGE IN AOFAS
1	38
2	28
3	34
4	45
5	16
6	17
7	37
8	50
9	38
10	42
11	17
12	9
13	37
14	33
15	12
16	35
17	37
18	26
19	18

Table3: AOFAS Scores and Radiographic (Kellgren-Lawrence Arthritis Grading Scale) Scores Pre and Post-Operatively

PATIENT	Pre AOFAS	Pre XR MTJ	Pre XR AJ	Post AOFAS	Post XR MTJ	Post XR AJ
1	43	0	0	81	0	0
2	50	0	0	78	0	0
3	51	1	0	85	1	1
4	45	0	1	90	0	1
5	55	0	0	71	0	0
6	55	0	0	72	0	0
7	48	0	0	85	0	0
8	40	0	1	90	0	1
9	41	1	1	79	1	1
10	33	0	1	75	0	1
11	51	0	1	68	0	2
12	62	0	0	71	0	1
13	55	1	0	92	1	0
14	48	0	0	81	0	0
15	50	0	1	62	0	1
16	31	0	0	66	1	0
17	55	0	0	92	0	0
18	47	1	0	73	1	0
19	46	0	0	74	1	0

	PRE	POST	AVG CHANGE
AVERAGE AOFAS	47.68	78.16	30.74
Stand Dev	7.75	9.07	11.58

Table 4: Postoperative SF-36 Scores

Patient	PF	RP	BP	GH	VT	SF	RE	HM
1	80	70	74	87	80	87.5	100	96
2	50	75	64	62	65	87.5	100	88
3	85	75	84	87	80	100	100	72
4	95	100	80	95	85	100	100	80
5	60	100	41	82	50	87.5	100	68
6	60	100	41	82	50	87.5	100	68
7	80	50	74	82	80	87.5	100	96
8	85	80	84	77	80	100	100	92
9	75	80	74	87	80	87.5	100	96
10	80	75	62	87	100	100	100	92
11	90	100	74	67	70	87.5	100	80
12	55	0	51	47	25	62.5	100	56
13	100	100	84	95	100	100	100	100
14	95	75	74	95	95	100	100	100
15	55	0	74	77	80	100	100	92
16	85	100	74	87	80	87.5	100	96
17	100	100	84	95	100	100	100	100
18	75	75	62	87	55	100	100	72
19	65	75	62	67	65	75	66.7	80
AVG SF-36	77.37	75.26	69.32	81.32	74.74	91.45	98.25	85.47
Stand Dev	15.84	30.11	13.45	12.78	19.61	10.25	7.64	13.21

A summary of case series and retrospective reviews on isolated subtalar fusions for adult acquired flatfoot is seen in Table 1.

While there was been a trend in the last decade for surgeons to opt for joint preservation procedure for correction of adult acquired flatfoot deformity, one cannot deny the time tested results and evidence of isolated subtalar arthrodesis. Many contrarians would argue theories such as exacerbation of adjacent joint arthritis, increased hindfoot rigidity, and need for revisional procedures due to pain or nonunion. The latter theory has not been supported by literature review, as the union rate seen from numerous case series showing greater than 95% union rate. Loss of follow up in many of these case series could very well be from unsatisfied patients, potentially skewing and underestimating nonunions.

While adjacent joint arthritis may be a factor, many of these patients are asymptomatic and cause and effect can often not be proven. There is only mild sacrifice of overall hindfoot motion when the subtalar joint is fused[26]. The joint-preserving anterior calcaneal osteotomy had encouraging results when utilized for children; however, this led to degenerative arthrosis of the calcaneocuboid joint when applied to adults[27]. Many experts on this topic subscribe to joint preservation osteotomies, but state these procedures are contraindicated in patients with continuous lateral foot pain, subtalar degenerative changes, or obesity. In patients with late stage II posterior tibial tendon dysfunction, many of these variables are already present[28]. Juxtapose these findings with the 85% satisfaction rate at five years with isolated subtalar arthrodesis[29].

Subtalar arthrodesis is a time tested technique that has shown great success since it was first introduced by Grice and Gallie as early as the 1940s[30, 31]. It can be employed for a wide variety of pathology, but most notably has shown success and positive outcomes in adult acquired flatfoot deformity.

This retrospective study looked at clinical and radiographic improvement of 18 patients (19 limbs) undergoing an isolated subtalar joint arthrodesis performed by the senior author. Twelve patients (13 limbs) underwent ancillary soft tissue procedures based upon on their symptomatology and deformity. All patients improved in AOFAS scores and the majority of patients (14/18 or 77.8%) stated they would undergo the same procedure again due to their improvement of activities and decreased lifestyle limitations.

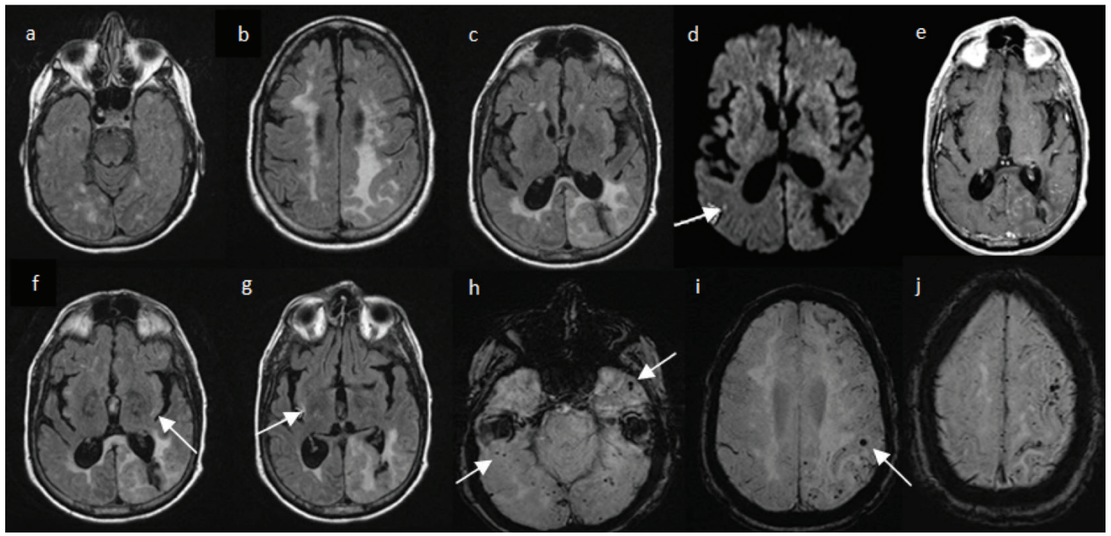
When comparing the mean postoperative AOFAS score of 78 to other mentioned studies, it was noted to be higher in most cases. The SF-36 scores were compared to population norms established in 1998[32]. This study's population had slightly higher and comparable scores when compared to this data. No patient experienced complete resolution of pain (0/10) following complete healing of their procedure; however, pain in most was limited to certain activities and much improved from the preoperative state. Patients with no preoperative radiographic arthritic findings demonstrated no further changes on radiographic evaluation at their most recent mid-term follow up. In contrast, asymptomatic patients with preoperative radiographic arthritic findings became symptomatic post-operatively. There was also slight progression of radiographic findings according to the Kellgren-Lawrence Arthritis Grading Scale.

This study included inherent limitations including the length of the follow-up of the patients as a longer time period would be useful in determining the arthritic changes that may occur greater than seven years after the isolated subtalar joint fusion. In addition, this study did not have a control group and had a relatively low number of patients. Further studies with a greater number of patients with a longer follow-up would allow for a more detailed analysis of prognostic factors.

| continued on p.11

Atypical MRI findings in a patient with cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL): a diagnostic dilemma

By **A.M. Alfieri, S. Warhadpande, Department of Radiology, Wexner Medical Center, Ohio State University, Columbus, OH**



INTRODUCTION

Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is, as the name suggests, a rare genetic disorder that predisposes the affected individual to hemorrhagic or ischemic strokes. CADASIL is inherited in an autosomal dominant fashion and occurs due to a mutation in NOTCH-3, a transmembrane receptor protein expressed on vascular smooth muscle cells.[1]

Mutated NOTCH-3 accumulates intracellularly in vascular smooth muscle cells and results in the progressive destruction and fibrosis of small and medium-sized cerebral penetrating arteries.[2] The ensuing luminal narrowing produces ischemia and, eventually, multiple lacunar infarcts.[3] The four cardinal symptoms in CADASIL include ischemic stroke, dementia, migraine with aura, and mood disturbances.[4] The most common manifestation of CADASIL are transient ischemic accidents (TIA) and subcortical strokes, which occur between the ages of 40 and 60. TIAs and sub-cortical strokes are followed by cognitive dysfunction and dementia.3 Migraines with aura are also common and generally appear around the age of 30.1 Although not as common as the other symptoms of CADASIL, mood disturbances have been reported in up to 20% of cases.[5]

With such a vast array of clinical presentations combined with its relatively rare occurrence, CADASIL can often present a diagnostic difficulty to physicians. In this report, we will first present an atypical case of CADASIL and the challenges that were faced on the path to reaching the diagnosis. We will then review the literature regarding the MRI findings in patients with CADASIL so that these features can be better understood and applied in cases suggestive of CADASIL.

CASE REPORT

A 67-year-old man presented to the neurology service with a three-month history of cognitive decline, confusion, and short-term memory impairment. Past medical history was relevant for Hypergammaglobulinemia of Waldenstroms, atrial flutter, hypertension, a left occipital lobe hemorrhagic stroke in 2008, and a TIA in 2009.

The patient's mini-mental status exam score was 16/30 on presentation. Patient was alert but oriented to only person and place. Five-minute recall to the words, "Mr. Johnson, umbrella, tunnel" was 0/3. Language and fluency were intact, but his comprehension to commands was impaired. Both cranial nerve exam and motor exam were unremarkable. Deep tendon reflexes were +2 bilaterally throughout and plantar stimulation elicited down-going toes bilaterally. Sensation to light touch and cerebellar function were grossly intact. A brain magnetic resonance imaging (MRI) study was subsequently obtained.

The MRI results revealed areas of confluent white matter hyperintensity (WMH) in the frontal, parietal, and occipital lobes in a relatively symmetric distribution. However, there was general sparing of the temporal lobes with little evidence of WMH in this region. Diffusion weighted imaging revealed multiple punctuate infarcts within the right cerebellar hemisphere, right occipital lobe, and right parietal lobe.

Susceptibility weighted imaging (SWI) showed several punctuate intraparenchymal hemorrhages, which were primarily distributed through the subcortical areas of both cerebral hemispheres. T1 films with contrast demonstrated some left parieto-occipital cortical swelling with adjacent signal abnormality, which was thought to be representative of edema and pial vessel engorgement. The patient had a prior stroke in this region (left occipital lobe) in 2008, so it is possible these findings may be related to the prior stroke.

These atypical MRI findings did not hint at a possible etiology and the differential included everything from CADASIL to amyloid angiopathy.

An extensive work-up followed, which included a bone marrow biopsy, an abdominal fat pad punch biopsy, and a skin biopsy. The patient was also scheduled for a brain biopsy when the skin biopsy results returned and revealed intracellular granular osmiophilic deposits within vascular smooth muscle cells, a finding pathognomonic for CADASIL.

During the hospital stay, the patient also suffered from two generalized tonic-clonic seizures. EEG monitoring revealed that the patient had been having almost continuous subclinical seizures. It was hypothesized by the care team that the 3-month history of cognitive decline and decreased mentation prior to the hospital admission was a result of subclinical seizure activity. Given the genetic implications of the CADASIL diagnosis, the patient's family was counseled and referred to a neuro-geneticist. Once the patient's seizures were confirmed to be under control, the patient was discharged.

DISCUSSION

This case represents the diagnostic challenge that often accompanies CADASIL. Imaging has been an important aspect of recognizing and differentiating CADASIL since it became recognized in 1990s.[6] Several radiographic findings have been described in CADASIL, though some are more specific to the disease than others. Since CADASIL can often masquerade as other diagnoses, such as MS[7] or encephalitis[8], it is important to understand the MRI characteristics of CADASIL so that it can be included in the differential when appropriate. This would allow for more timely acquisition of appropriate diagnostic tests, including genetic testing and/or skin biopsy, and would prevent inappropriate treatment courses from being pursued.

In the case of our patient, the MRI features were atypical, with sparing of the temporal white matter, so the diagnosis of CADASIL was not entertained until several other diagnostic studies had been completed. This illustrates the important point that, although understanding the MRI findings in CADASIL can be beneficial diagnostically, these features are not always present and hence CADASIL should not immediately be removed from the differential diagnosis in their absence if clinical suspicion or family history is suggestive. Regardless, having a thorough understanding of the radiographic features of CADASIL is important and the duration of this section will focus on these features.

Early imaging studies of CADASIL demonstrated the presence of white matter involvement as an important feature of the disease. Chabriat et al. assessed 75 patients with MRI and found that 47% of cases showed hypointensities on T1-WI that increased in frequency with age. T2-WI revealed that 90% of the patients had hyperintensities within the white matter (WMH) that was symmetric and was most frequent in the periventricular and deep white matter regions, but also present within the brainstem, basal ganglia, and external capsule of several patients. Interestingly, the severity of signal abnormality within the periventricular, basal ganglia, and infratentorial regions was higher in the symptomatic patients compared to the asymptomatic patients.[9] The involvement of WMH has been subsequently demonstrated in numerous studies of CADASIL confirmed patients. [10,11,12]

While WMH is a prominent finding in MR images of CADASIL patients, it can also be found in other conditions such as MS, subcortical arteriosclerotic encephalopathy (sSAE), and ischemic leukoaraiosis.[7,10,13] Thus several studies began searching for MRI findings that were more specific to CADASIL. O'Sullivan et al. compared MRI studies from 20 patients with CADASIL to 20 patients with leukoaraiosis. The results showed that both groups had marked WMH involvement of the periventricular, frontal and parietal regions, but that on T2-WI hyperintensity of the temporal pole and external capsule-insula region was significantly greater in CADASIL.[13]

Similarly, Auer et al. found significantly higher T2-WI and FLAIR hyperintensity in the temporal white matter in CADASIL patients compared to patients with sporadic subcortical arteriosclerotic encephalopathy.[10] Several other studies have also demonstrated temporal lobe WMH as an important MRI characteristic for identifying CADASIL.[11,12,14,15] In our patient, the finding of temporal lobe sparing was atypical and initially led us down a different diagnostic path. Thus, although temporal lobe WMH are frequently present in CADASIL, it is important to remember that this finding is not universal and CADASIL should still be considered if other findings are suggestive.

Lacunar infarcts/lesions are further MRI characteristics that have been identified as potential markers for CADASIL. Van den Boom et al. found that subcortical lacunar lesions (SLL) on MRI studies were 59% sensitive and 100% specific for CADASIL compared to healthy controls, elderly patients with vascular risk factors, patients from CADASIL families who were negative for the NOTCH 3 mutation, and patients with small vessel disease with WMH.[16] When present, the SLLs were found in the anterior temporal horn in all cases and in the frontal lobe in 25% of cases. A subsequent microscopic exam of one of the CADASIL patients revealed the SLLs to be caused by distension of the perivascular space of the perforating arteries at the grey-white junction.[16]

A subsequent study by the group also found evidence of SLLs in 56% of CADASIL patients ages 41-50 with the value increasing to 73% in CADASIL patients between the ages of 51-60.[12] Although lacunar lesions and infarcts are often not separate en-

Atypical MRI

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tities in the literature, this group differentiated the two based on location with SLLs being at the grey-white junction and lacunar infarcts located in regions that did not extend to cortical gray matter. Thus along with the findings for SLLs, they also found lacunar infarcts within the supratentorial white matter and basal ganglia in 75% of CADASIL patients between 31-40 and in over 90% of CADASIL patients >40 years of age.[12] The presence of lacunar infarcts in CADASIL has since been confirmed by several other studies.[17,18,19]

One final radiographic feature that has been identified in CADASIL is the presence of microbleeds. Dichgans et al. found evidence of microbleeds in 69% of patients with CADASIL compared to 0% of age matched control subjects on gradient echo T2 MRI. The microhemorrhages were found in a variety of areas throughout the brain, with the cortical-subcortical region being most frequent, but importantly were largely outside the areas of hyperintensity on T2/PD-WI.[20] Microhemorrhages were also demonstrated in 19% from ages 41-50 and 47% of patients > 50 years of age in another study, suggesting they are a later feature of the disease.[12]

It has subsequently been demonstrated that the number of cerebral microhemorrhages in CADASIL patients was related to the development of intracerebral hemorrhages.[21] Since the microbleeds in CADASIL occur in areas distinct from the ischemic lesions and WMH, it is probable that microhemorrhage and infarction are independent issues in the pathogenesis of CADASIL. This conclusion is supported by Liem et al. who found that microbleed load on MR at baseline was only predictive of microbleed load at the seven-year follow up, while WMH and lacunar infarcts on MR were each predictive of both lacunar infarct load and WMH load at the seven-year follow up.[22]

Apart from their utility in aiding diagnosis of CADASIL, MRI features of CADASIL have also been shown to be correlated with clinical features of the disease. In a cohort study of 112 patients with CADASIL, Singhal et al. found that a history of stroke correlated with total WMH score, along with internal capsule WMH and pontine WMH involvement independently. They also found that presence subcortical white matter hyperintensity correlated with presence of psychiatric disorders and with presence of dementia.[11]

Several other studies have also examined the MR findings in CADASIL, but with the focus on their relationship to cognition. Peters et al. found that brain atrophy on MR was significantly correlated with disability and global cognitive performance. [23] Similarly, Jouvent et al. found that the degree of brain atrophy was significantly associated with cognitive defects and disability in 147 CADASIL patients, but also that the degree of brain atrophy was correlated with the volume of lacunar lesions and mean apparent diffusion coefficient.[24] Several other studies have also identified lacunar infarcts on MRI as being correlated with cognitive impairment. [17,25, 26] Taken together these findings suggest that lacunar lesions may lead to cognitive decline or dysfunction through inducing structural changes in the brain leading to brain atrophy.

In conclusions, it is important for clinicians to keep CADASIL in mind when a patient's MRI demonstrates symmetric WMH, especially in the anterior temporal lobe and external capsule, subcortical lacunar infarcts, and/or diffuse microhemorrhages. However, as in this patient, it is important to remember that these findings are not absolute and CADASIL should not be excluded from the differential diagnosis if clinical suspicion is high. Finally, as correlations between MRI findings in CADASIL and clinical features of the disease have been found, imaging may become an important tool for monitoring progression and predicting clinical course in the future.

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Isolated Subtalar Joint Arthrodesis

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Table 1: Summary of Literature Review

AUTHOR	FUSIONS	AAFD PATIENTS	TECHNIQUE	F/U (MONTHS)	RESULTS	UNION RATE
Easley et al[7]	184	0	1-2 screw with bone graft in 145 feet	51	AOFAS improved 24 to 70	84%
Mann et al[6]	44	11	One screw	59.5	AOFAS 89 at final follow up and 93% satisfaction	100%
Chiang et al[12]	15	0	Two screws with femoral head allograft	36	AOFAS and VAS improved with 93.3% satisfaction	100%
Garras et al[13]	24	0	Two screws with platelet-rich plasma and femoral head allograft	35.8	AOFAS improved 21 to 71	90%
Trnka et al[14]	41	0	1-2 screws with femoral head allograft or iliac autograft	70	AOFAS improved 21.1 to 68.9 with 97% satisfaction	87%
Davies et al[22]	95	0	One screw	N/A	93% had good to fair outcome	95%
Yu et al[15]	48	48	One screw with additional point(s) of fixation: Steinmann pin, K-wire, screw.	13.05	Improvement in radiographic parameters	n/a
Russotti et al[19]	45	15	One screw or steinmann pin with iliac crest	57	Excellent results in 87% with 90% satisfaction. No secondary degenerative joint disease of adjacent joints	98%
Fellmann et al[20]	36	12	One steinmann pin along with iliac crest and OP-1 in 6 cases	32.5	VAS improved 4.4 to 1.1 with 78% good to excellent result. Range of motion to ankle and Chopart's joint maintained	95%
Joveniaux et al[21]	28	2	1-2 staples with iliac autograft, cancellous autograft, or bone substitute	56	AOFAS improved 49 to 76.5. Asymptomatic mild arthritic changes adjacent joints in 43-65%	100%
Dennyson et al[18]	48	48	One screw with iliac autograft	36	90% had satisfactory correction of deformity. Higher rate of nonunion when casted less than 8 weeks	94%
Kitaoka et al[16]	21	21	One screw and without bone graft	36	76% had excellent to good result with improvement in radiographic parameters	100%
Haskell[23]	101	18	One screw autograft from sinus tarsi and anterior process	N/A	.Fixation removed in 13% with average time to fusion mean 12.3 +/- 3.4 weeks	98%
Catanzariti et al[25]	40	19	Two screws with various bone graft in 45% of cases	15	80% were satisfied and 83% would undergo procedure again. Minor complication rate 55%	90%
Mangone et al[24]	34	11	One screw	30.8	83% would undergo procedure again with 100% improvement in 45% of patients.	100%
Johnson et al[17]	17	17	One screw with autogenous iliac crest or distal tibia. Flexor digitorum longus tendon transfer with spring ligament repair also performed.	27	AOFAS and Maryland score improved to 82 and 86 respectively with improvement in radiographic parameters	100%

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An observational study for detection of ICU Delirium and Delirium subtypes at St. Vincent Charity Medical Center

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ABSTRACT

Background

Several studies suggested that delirium in Intensive Care Unit (ICU) setting is under-recognized and under-documented by both physicians and nursing staff. This observational study compares rates of detection of ICU delirium using current standard of care at St. Vincent Charity Medical Center (SVCMC) to the standardized tool confusion assessment method (CAM-ICU) and evaluates some of the risk factors of delirium at SVCMC ICU.

Objective

To compare rate of detection of delirium in ICU using current standard of care at SVCMC and standardized tool CAM-ICU. To hypothesize that the rate of undetected delirium & delirium subtypes in ICU patients is higher and we can improve its detection by utilizing CAM-ICU forms among residents and nursing staff. The secondary objective was to evaluate some of the risk factors of delirium at SVCMC ICU.

Study Design and Method

We conducted an observational study where 100 patients were assessed by CAM-ICU method if they spent at least 48 hours in the ICU. They were seen twice a day for at least three consecutive days or until delirium was detected. We reviewed daily assessment notes of residents and nurses and compared them with our findings. The kappa statistics was done to measure agreement over and beyond chance agreement.

Results

For the agreement comparisons, each of the intake assessments from researchers and residents as well as researchers and nurses, the value of kappa was found to be 0.18 (95% CI: 0.02 – 0.33). The three-way agreement of researchers, nurses and residents assessment also had kappa value of 0.18 (95% CI: 0.02 – 0.33). This agreement is considered "poor" agreement by the traditional Landis & Koch way of characterizing kappa. By using kappa statistics, we found that there is significantly more agreement than by chance alone ($p = 0.002$). So, there is better than chance agreement, but it is "poor" agreement.

The prevalence of delirium was 30% (30/100). Among them, study confirmed higher rates of undetected hypoactive delirium 66.6% (20/30 patients), and 30% (9/30) for mixed delirium. Delirium was observed in 30% of patients within the first five days of ICU admission. The mean age was 68+/-14 in patients with delirium and 65+/-11.6 in patients without delirium. Also, delirious patients had increased hospital length of stay (LOS)(median LOS 10 days) versus non-delirious patients(median LOS 8 days) ($p < 0.0001$). Delirious patients had higher SAPS II score 31.3 +/- 13.8 versus 24.5 +/- 10.6 for non-delirious patients ($p < 0.0001$). Delirious patients also had higher numbers of TLC and Foley's catheterization ($p < 0.05$) than non-delirious patients.

Conclusion

CAM-ICU is an excellent screening tool for early diagnosis of ICU delirium and should be integrated in daily residents and nurses' assessment of patients to increase detection rate of delirium as early as within 2-5 days of ICU admission.

INTRODUCTION

Delirium is characterized by acute onset and fluctuating course of inattention. It is a change in cognition or the development of a perceptual disturbance that is not better accounted for by a preexisting, established, or evolving dementia. The disturbance develops over a short period of time (usually hours to days) and tends to fluctuate during the course of the day [1].

In delirium, patient's ability to receive, process, store, and recall information is impaired. This state is usually reversible and is a direct consequence of a medical condition, substance intoxication or withdrawal, use of a medication, toxin exposure, or a combination of these factors.

Delirium has an enormous impact upon the health of elderly patients. Patients with delirium experience higher mortality, prolonged hospitalizations, functional and cognitive decline and higher risk for institutionalization. Signs of delirium may persist longer than a year, especially in patients with underlying dementia. Even though, delirium can be reversible, it can create future problems for fragile, elderly patients.

There are three subtypes of delirium: hyperactive, hypoactive and mixed. Hyperactive delirium is characterized by agitation, restlessness and attempts to remove tubes and lines. Hypoactive delirium is characterized by withdrawal, flat affect, apathy, lethargy and decreased responsiveness. Mixed delirium is considered when patient fluctuates between hyperactive and hypoactive delirium. Hypoactive and mixed delirium are more common in ICU setting and are often unrecognized. Multiple assessment tools are available to detect delirium in patients and one of them is CAM-ICU[2, 3].

OBJECTIVES

1. To compare the rate of detection of delirium in ICU patients using current standard of care at SVCMC versus the standardized tool CAM-ICU.
2. To hypothesize that the rate of undetected delirium & delirium subtypes in ICU patients is higher and that we can improve its detection by utilizing CAM-ICU forms among residents and nursing staff.
3. To evaluate risk factors of delirium at SVCMC ICU.

METHODS AND PROCEDURES

Study design: Observational study with bedside evaluation and documentation review.

Setting: St. Vincent Charity Medical Center (SVCMC), an urban community teaching hospital in Cleveland, Ohio.

Participants: 100 patients admitted or transferred to ICU.

RASS and CAM-ICU Worksheet

Step One: Sedation Assessment

The Richmond Agitation and Sedation Scale: The RASS*

Score	Term	Description	
+4	Combative	Overtly combative, violent, immediate danger to staff	
+3	Very agitated	Pulls or removes tube(s) or catheter(s); aggressive	
+2	Agitated	Frequent non-purposeful movement, fights ventilator	
+1	Restless	Anxious but movements not aggressive vigorous	
0	Alert and calm		
-1	Drowsy	Not fully alert, but has sustained awakening (eye-opening/eye contact) to *voice* **(≥10 seconds)**	Verbal Stimulation
-2	Light sedation	Briefly awakens with eye contact to *voice* **(<10 seconds)**	Verbal Stimulation
-3	Moderate sedation	Movement or eye opening to *voice* **(but no eye contact)**	Verbal Stimulation
-4	Deep sedation	No response to voice, but movement or eye opening to *physical* stimulation	Physical Stimulation
-5	Unarousable	No response to *voice or physical* stimulation	Physical Stimulation

Procedure for RASS Assessment

1. **Observe patient**
   a. Patient is alert, restless, or agitated. **(score 0 to +4)**
2. **If not alert, state patient's name and *say* to open eyes and look at speaker.**
   a. Patient awakens with sustained eye opening and eye contact. **(score –1)**
   b. Patient awakens with eye opening and eye contact, but not sustained. **(score –2)**
   c. Patient has any movement in response to voice but no eye contact. **(score –3)**
3. **When no response to verbal stimulation, physically stimulate patient by shaking shoulder and/or rubbing sternum.**
   a. Patient has any movement to physical stimulation. **(score –4)**
   b. Patient has no response to any stimulation. **(score –5)**

If RASS is -4 or -5, then **Stop** and **Reassess** patient at later time
If RASS is above - 4 (-3 through +4) then **Proceed to Step 2**

*Sessler, et al. AJRCCM 2002; 166:1338-1344. Ely, et al. JAMA 2003; 289:2983-2991.

Step Two: Delirium Assessment

Feature 1: Acute onset of mental status changes or a fluctuating course

And

Feature 2: Inattention

And

Feature 3: Disorganized Thinking OR **Feature 4**: Altered Level of Consciousness

= DELIRIUM

Inclusion Criteria: 18 years and older, patients admitted to the ICU and CSU for at least 48 hours.

Exclusion Criteria: Younger than 18 years old, non-English speaking patients, admission for an aphasic stroke.

We assessed 100 Intensive Critical Care patients 48 hours after their admission or transfer to the Unit. These patients were evaluated twice a day for maximum of three days or until they were found to have delirium. By using CAM-ICU checklist[2, 3, 4], we tried to detect and diagnose different forms of delirium. At the same time, we reviewed progress notes of primary residents and nurses regarding their assessment of delirium. To assure valid CAM-ICU results, we assessed patient's sedation status by utilizing RASS score[5, 6]. The RASS score is a 10-point scale for evaluation of the level of sedation. A patient with a RASS score of -3 to +4 was considered eligible to be assessed by the CAM-ICU to determine the presence of delirium.

The Confusion Assessment Method (CAM) was created in 1990. CAM-ICU is an adaptation of this tool for use in ICU patients. The CAM-ICU score is determined by examining patient for inattention, acute and fluctuating changes in mental status, disorganized or incoherent thinking, and altered level of consciousness. The CAM-ICU has been developed, validated and applied into ICU settings and multiple investigators have used the same method to identify patients with delirium. Delirium is defined in terms of four diagnostic features, and deemed present when a patient has positive Feature 1 and Feature 2 and either Feature 3 or 4 (see CAM-ICU schematic).

DATA COLLECTION

All of the researchers reviewed the training manual and videos from the icudelirium.org website.

After obtaining verbal consent, we assessed patient's acute change of mental status evidenced by fluctuation on a sedation scale (i.e. RASS), GCS, or previous delirium assessment. To determine baseline mental status, we used information from family or friends, nursing home staff, and History and Physical. For example, if the patient was young and was admitted from home with no documented neuro-cognitive impairment or history of stroke, then we assumed that the patient had a "normal" baseline mental status. If the patient was older, had documentation of a stroke or dementia, or came from a nursing home, then we probed family or the institution for more information on the patient's pre-hospital baseline mental status.

We assessed inattention by asking patient to squeeze our hand whenever they heard the letter "A" in the series of 10 letters. Errors were counted when patient failed to squeeze on the indicated absence of delirium. If acute change of mental status was absent and the number of errors was between 0 – 2, CAM-ICU was considered negative for delirium. Patients were reevaluated on a daily basis until they were transferred to RNF (up to 48 hours post transfer) for maximum of three days. If acute

change of mental status was present and the number of errors were 0 – 2, we used RASS score to measure level of consciousness.

For disorganized thinking assessment we asked yes/no questions. For example:

1. Will a stone float on water?
2. Are there fish in the sea?
3. Does one pound weigh more than two pounds?
4. Can you use a hammer to pound a nail?

Errors were counted when the patient incorrectly answered a question. If a patient answered the four questions correctly we still assessed the command.

"Hold up these many fingers" (Hold 2 fingers in front of patient)

"Now do the same thing with the other hand" (Do not repeat number of fingers).

Combined number of > 1 would be recorded.

Overall CAM-ICU scoring: Feature 1 plus 2 and either 3 or 4 present = CAM-ICU positive[3, 4].

We reviewed residents and nursing staff notes for presence of delirium documentation. Additional data was collected from EMR (Meditech), such as demographics, length of stay, reason for admission, list of medications[7, 8], use of physical restraints, presence of IV lines, urinary catheter and other related co-morbidities[9, 10, 11]. We calculated SAPS II score (simplified acute physiology score). It is a severity score and mortality estimation tool developed from a large sample of medical and surgical patients in North America and Europe[12].

Table 1: Interrater detection of delirium by researchers (CAMICU* tool) vs. Residents and Nurses

Rater	k (95% CI)
Researchers vs. Residents	0.18 (0.02-0.33)
Researchers vs Nurses	0.18 (0.02-0.33)
Researchers vs. Nurses vs. Residents (three way agreement)	0.18 (0.02-0.33)

*CAM ICU -- Confusion Assessment Method for the Intensive Care Unit

(p=0.002) for all comparisons so there is better than chance agreement for all of them

Table 2: Patient characteristics

Characteristics	Frequency (total n=100)
Age, mean ± SD	67 ± 13.3
- Patients with delirium	68 ± 14
- Patients without delirium	65 ± 11.6
Gender	
Male, %	54
- Male with delirium	12
Female, %	46
- Female with delirium	18
SAPS II Score*, mean ± SD	26.73 ± 11.95
- Patients with delirium mean ± SD	31.3 ± 13.8
- Patients without delirium mean ± SD	24.5 ± 10.6
ICU* admission diagnosis, n (%)	
Acute respiratory distress syndrome	9
Myocardial infarction or arrhythmia	15
Congestive heart failure	5
Hepatic or renal failure	8
Chronic obstructive pulmonary disease	9
Gastrointestinal bleeding	4
Malignancy	2
Drug overdose	4
CABG*	8
Sepsis or Septic shock	18
Hypertensive Emergency	4
Bariatric surgery	5
General surgery/Orthopedic surgery	9

*ICU -- Intensive Care Unit,
*SAPS II Score -- Simplified Acute Physiology Score
*CABG --Coronary Artery Bypass Grafting

DATA STORAGE AND CONFIDENTIALITY

Data was stored in the form of encrypted Excel spreadsheets to which only the primary investigators had access. There were no patient identifiers. Patient medical records were retained for the same period according to the document retention and disposition schedule of St. Vincent Charity Medical Center.

STATISTICAL METHODS

The kappa statistic (Cohen's kappa) was used to measure agreement over and beyond chance agreement. The two-way agreement comparisons of researcher and resident assessment of patients as well as researcher and nurses assessment along with three-way agreement of researcher, nurse and resident assessment were analyzed and kappa value were estimated. For evaluation of presence of risk factors in delirious patients, fisher's test was completed to determine p value. A p-value less than 0.05 was considered to be statistically significant.

RESULTS

A total of 100 patients were assessed at bedside. 30 patients (30%) were found to be delirious by using CAM-ICU tool. Among them, 20 patients (66.6%) had hypoactive delirium, 1 patient (3.33%) has hyperactive delirium, and 9 patients (30%) had mixed delirium. For the agreement comparisons, each of researchers & resident assessment, as well as researchers and resident assessment, the value of kappa was 0.18 (95% CI: 0.02 – 0.33). The three-way agreement of researchers, residents and nurses assessment also had kappa value of 0.18 (95% CI: 0.02 – 0.33).

This agreement is considered "poor" agreement by the traditional Landis & Koch way of characterizing kappa. For the kappa statistic there is a statistical hypothesis test of whether there is significantly more agreement than one would find by chance alone (i.e., a test as to whether the true value of kappa is zero), whereby it is found that there is significantly more agreement than by chance alone (p = 0.002). So, there is better than chance agreement, but it is "poor" agreement (Table 1). For completeness, we also examined the agreement of nurses and residents assessments and found for the kappa statistic that kappa = -0.04 (95% CI: -0.07 – -0.01). This is actually less agreement than what one could expect by chance alone and the corresponding p-value (p=0.68) is not statistically significant.

Table 2 illustrates the demographic values of our study patients.

In this observational study, out of 100 patients, 30 had delirium. The prevalence of delirium was 30%. Among them, our study confirmed higher rates of undetected hypoactive delirium as 66.6% (20/30) patients, and 30% (9/30) of mixed delirium (Figure 1 & 2). Delirium was observed in 30% of patients within the first 5 days of ICU admission. The mean age of patient was 67 13.3 (68 14 in patients with delirium and 65 11.6 in patients without delirium).

There was a significant relationship between increased length of hospital stay in patients with delirium (median length of stay 10 days) versus non-delirious patients (median length of stay 8 days) (p < 0.0001) (Figure 4). The mean SAPS II score was 26.73 11.95. Among patients with delirium SAPS II score was 31.3 13.8 and it was 24.5 10.6 in patient without delirium (p < 0.0001) (Figure 5).

We also found that patients with delirium had higher numbers of TLC and Foley's catheterization (p < 0.05) (Figure 6 & 7).

There was also a significant relationship between higher use of benzodiazepine in delirious patients v/s non-delirious patients (p = 0.0009) (Figure 8).

We found no significant relationship between use of antipsychotics and delirium (p = 0.1228) (Figure 9) and the use of opiates and delirium (p=0.8216) (Figure 10).

DISCUSSION

Several studies suggested that delirium in ICU setting is under-recognized and under-documented by both physicians and nursing staff[13, 14, 15]. Under-documentation of delirium in the medical record is supported by our findings, in that there was no chart documentation for 30% (30/100) of delirium cases identified by the CAM-ICU.

There are three subtypes of delirium: hypoactive, hyperactive and mixed[16, 17, 18]. Hyperactive delirium is characterized by agitation, restlessness, and attempts to remove tubes and lines. Hypoactive delirium is characterized by withdrawal, flat affects, apathy, lethargy, and decreased responsiveness. Mixed delirium is when patients fluctuate between the two. In ICU patients, mixed and hypoactive forms of delirium are often unrecognized without routine monitoring. Our study confirmed higher rates of undetected hypoactive as 66.6% (20/30) patients, and 30% (9/30) of mixed delirium. Hypoactive delirium generally has worse prognosis and missed without active monitoring (4). Early detection and treatment can lower ICU, hospital and 30-day mortality[19, 20, 21, 22, 23]. It also decreases hospital-acquired complications related to delirium and shortens hospital length of stay.

A number of methods are available to detect delirium in critically ill patients. There is evidence that even ICU physicians recognize less than one third of delirious critically ill patients without using assessment tool for detection of delirium to aid in their diagnosis[24]. There are several validated instruments to assess delirium, including Cognitive Test for Delirium, abbreviated Cognitive Test for Delirium, Confusion Assessment Method for the Intensive Care Unit (CAM-ICU), Intensive Care Delirium Screening Checklist, Neelon and Champagne Confusion Scale, and the Delirium Detection Score[25]. The most extensively studied instrument is the CAM-ICU, which was validated to assess delirium at the bedside in nonverbal ventilated ICU patients[26]. Using a structured format, this tool evaluates four features, namely, acute onset or fluctuating course, inattention, disorganized thinking, and altered level of consciousness. Two recent systematic reviews pooled several studies evaluating the accuracy of CAM-ICU[27, 28]. The majority of the studies included in the systematic reviews showed that the CAM-ICU is a highly accurate instrument for the diagnosis of delirium in the ICU.

CONCLUSION

Delirium is a frequent complication in critical care patients. Delirium in ICU patients is a dynamic and complex process. CAM-ICU tool can be helpful in detection of ICU delirium and should be integrated in the daily residents and nurses assessments to increase detection rate of delirium. The rate of detection of delirium in ICU using current standard of care at SVMC was lower compared to standardized tool CAM-ICU. In summary, our study suggests that formal training and implementation of daily delirium assessment

CAM-ICU Worksheet

	Positive	Negative
Feature 1: Acute Onset or Fluctuating Course Positive if you answer 'yes' to either 1A or 1B.		
1A: Is the pt different than his/her baseline mental status? Or **1B:** Has the patient had any fluctuation in mental status in the past 24 hours as evidenced by fluctuation on a sedation scale (e.g. RASS), GCS, or previous delirium assessment?	**Yes**	**No**
Feature 2: Inattention Positive if either score for 2A or 2B is less than 8. Attempt the ASE letters first. If pt is able to perform this test and the score is clear, record this score and move to Feature 3. If pt is unable to perform this test or the score is unclear, then perform the ASE Pictures. If you perform both tests, use the ASE Pictures' results to score the Feature.	**Positive**	**Negative**
2A: ASE Letters: record score (enter NT for not tested) Directions: Say to the patient, *"I am going to read you a series of 10 letters. Whenever you hear the letter 'A,' indicate by squeezing my hand."* Read letters from the following letter list in a normal tone. S A V E A H A A R T Scoring: Errors are counted when patient fails to squeeze on the letter "A" and when the patient squeezes on any letter other than "A."	**Score (out of 10): _____**	
2B: ASE Pictures: record score (enter NT for not tested) Directions are included on the picture packets.	**Score (out of 10): _____**	
Feature 3:Disorganized Thinking Positive if the combined score is less than 4	**Positive**	**Negative**
3A: Yes/No Questions (Use either Set A or Set B, alternate on consecutive days if necessary): **Set A** 1. Will a stone float on water? 2. Are there fish in the sea? 3. Does one pound weigh more than two pounds? 4. Can you use a hammer to pound a nail? **Set B** 1. Will a leaf float on water? 2. Are there elephants in the sea? 3. Do two pounds weigh more than one pound? 4. Can you use a hammer to cut wood? **Score** ___(Patient earns 1 point for each correct answer out of 4) **3B:Command** Say to patient: "Hold up this many fingers" (Examiner holds two fingers in front of patient) "Now do the same thing with the other hand" (Not repeating the number of fingers). *If pt is unable to move both arms, for the second part of the command ask patient "Add one more finger) **Score**___(Patient earns 1 point if able to successfully complete the entire command)	**Combined Score (3A+3B): _____ (out of 5)**	
Feature 4: Altered Level of Consciousness Positive if the Actual RASS score is anything other than "0" (zero)	**Positive**	**Negative**
Overall CAM-ICU (Features 1 and 2 and either Feature 3 or 4):	**Positive**	**Negative**

ICU Delirium

(cont. from previous page)

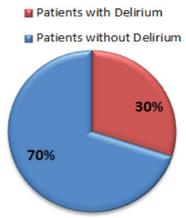

Figure 1

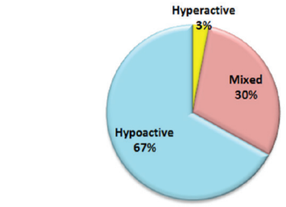

Figure 2: Subtypes of Delirium

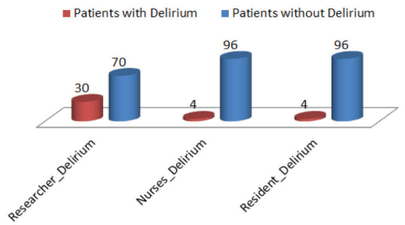

Figure 3: Detection of Delirium

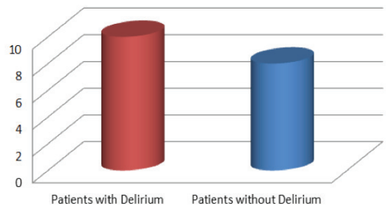

Figure 4: Length of Stay - Median (p value <0.0001)

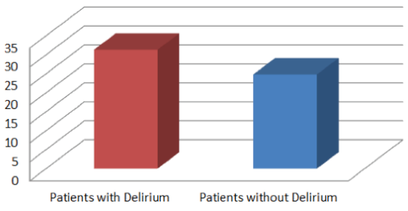

Figure 5: Average SAPS II Score (p value <0.0001)

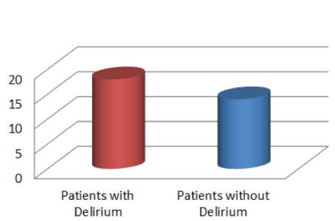

Figure 6: TLC & Delirium (p value = 0.002)

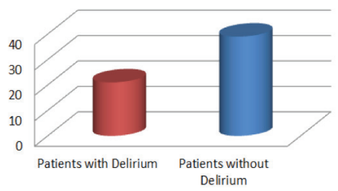

Figure 7: Foley & Delirium (p value <0.05)

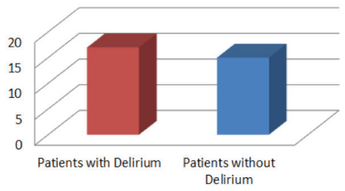

Figure 8: Benzodiazepine & Delirium (p value = 0.0009)

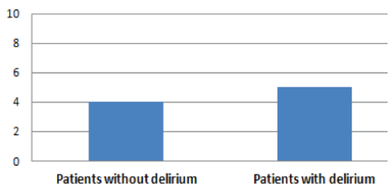

Figure 9: Antipsychotic & Delirium (p value = 0.1228)

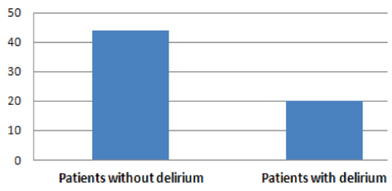

Figure 10: Opiates & Delirium (p value = 0.8216)

by using CAM-ICU should be attempted. Because delirium is a fluctuating disorder, and single daily observation can substantially underestimate the prevalence of delirium, the ICU nurses and residents should be trained to administer the instrument on each shift along with assessments of sedation and acuity.

Residents should re-evaluate the need for restraints and bladder catheters daily. They should also evaluate the use of sedatives (e.g. benzodiazepines or opiates) and medications with anti-cholinergic activity, and consider discontinuation if not required.

Overall, our data suggest that CAM-ICU is an excellent screening tool for the diagnosis of delirium in critically ill patients.

LIMITATIONS

This is an observational study. We acknowledge possibility of delirium development in some patients several days after we completed our assessments. Focus of this study was to detect delirium within five days of admission and did not extend evaluation after that time period. There is non-homogenous gender distribution in the delirium subgroup.

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Anti-synthetase syndrome

(cont. from p 3)

to skilled nursing facility for rehabilitation.

DISCUSSION

Anti-synthetase syndrome (ASS) is a systemic autoimmune syndrome characterized by the presence of anti-aminoacyl t RNA antibodies accompanied by a constellation of clinical findings, including polymyosistis-dermatomyositis (PM-DM), ILD, "mechanic hands" and Raynaud's phenomenon, etc. This is relatively uncommon clinical entity and is considered as a subset of idiopathic inflammatory myopathies.

ASS mostly affects adults, at any age, and is more common in women with a female to male ratio of 2:1. The clinical presentation of ASS varies greatly among the patients as does the severity of involvement of different organs. Among the anti-synthetase antibodies more frequently associated with lung involvement is the anti-Jo-1 antibody[1]. ILD is especially prevalent in ASS occurring in approximately 75% of patients with anti-Jo-1 antibody compared to 30% of patients with idiopathic inflammatory myopathies in the absence of antisynthetase antibodies.

It is noteworthy that, when present, the lung disease is the main determinant of survival in patients with ASS. Surgical lung biopsies in patients with ILD-associated ASS may show different histological features including nonspecific interstitial pneumonia (NSIP), usual interstitial pneumonia (UIP), cryptogenic organizing pneumonia (COP) or diffuse alveolar damage [2,3].

Cryptogenic pneumonia is now a well-recognized entity with characteristic clinical and radiological features and pathologic diagnostic criteria including the lack of identifiable cause such as drug-induced or infection-associated organizing pneumonia. Corticosteroids are the current standard of treatment of COP. Cyclophosphamide may be considered in severely ill patients who show no improvement with corticosteroid treatment within a few days, and in patients who fail to improve despite a prolonged course of corticosteroids[4].

ILD is the main cause of mortality and morbidity in inflammatory myopathies and ASS in particular. Our patient predominantly complained of respiratory illness, HRCT demonstrated consolidations consistent with acute ASS associated COP/ILD. This case uniquely demonstrates how the diagnosis of ASS may not be clinically obvious on initial presentation, but may appear upon further investigation. We need to consider broad differential diagnosis for suspected infectious pneumonia cases that are not responding to standard antibiotic regimens[5]. Early diagnosis and treatment can prevent disease progression and improve patient outcome.

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Iodide mumps:

A rare case of contrast induced sialadenitis after fistulogram in an end stage renal disease patient

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LEARNING OBJECTIVES

1. Iodine from iodinated contrast media may rarely elicit non inflammatory edema of the salivary glands. Acute sialadenitis following contrast administration is characterized by rapid, painless, bilateral enlargement of salivary glands.
2. Iodide mumps is usually a benign, selflimiting condition and may recur with further exposure to iodinated contrast.

INTRODUCTION

Iodide mumps or contrast induced acute sialadenitis is characterized by the rapid, painless enlargement of the parotid and submandibular glands following the use of iodinated compounds. T he pathogenesis of this adverse reaction remains unclear. It may be due to an idiosyncratic reaction or related to toxic accumulation of iodide in the ductal systems of the salivary glands.[1] The onset of symptoms can start within a few minutes to five days after contrast administration.[2] The course of iodineinduced sialadenitis is usually benign, and rapid resolution of symptoms is expected without definite treatment. The symptomatic management includes parenteral non steroidal antiinflammatory drugs (NSAIDs), steroids and dialysis.[3]

CASE

We report a case of 65 years old African American female, end stage renal disease on hemodialysis 3 times/ week, presented in the emergency room with complaint of facial andupper neck swelling, one day after fistulogram.

The past medical history was significant for increasing right arm swelling for last 23 weeks and inability to access right arm AV fistula for dialysis. A dialysis permacath was placed in left subclavian vein for access. A fistulogram was performed to check the patency of the AV fistula with visipaque® (Iodixanol), a nonionic iodine containing contrast media. It showed stenosis at the junction of right subclavian and superior vena cava (SVC). Balloon angioplasty was performed and repeat fistulogram showed good resolution of stenosis with less than 10% remaining. The patient was stable in PACU post procedure and went home without immediate complication.

The next day she woke up with lower facial and upper neck swelling. The patient did not have any signs of SVC obstruction including superficial venous prominence on chest, arm edema, cyanosis and plethoric face. She also denied any hoarseness of voice, difficulties in breathing but complained of pain during mouth opening. Nifedipine was stopped for suspicion of drug induced angioedema.

Bilateral tender submandibular glands were noted on examination (Images 1&2). CT of the neck and face showed e nlarged bilateral submandibular glands with infiltration of adjacent fat planes and diffuse soft tissue swelling in submandibular region and upper neck (Image 3&4, CT scan).

Image 1

Image 2

Images 1&2: Swelling in lower face and upper neck around the submandibular glands

Initial labs including CBCD, CMP, electrolytes and acidbase were essentially normal except elevated WBC count of 11,000 but no bandemia. BUN and Cr values were also high commensurate with ESRD. The patient was started on unasyn on day 1 for a possible infective cause of sialadenitis. It was stopped next day as there were no signs of infection including fever, chill, tachycardia and no pus discharge from submandibular duct.

A diagnosis of contrast induced sialadenitis was made and patient was started on IV ketorolac, decadron, fluid and pain management. Emergency intubation kit was prepared at bedside for possible airway compromise which was not required. Hemodialysis was done within 24 hours of admission and the submandibular swelling improved significantly in next 23 days. The patient was discharged in a stable condition on oral tapering dose of prednisone for 2 weeks.

DISCUSSION

Visipaque® (Iodixanol) is a nonionic iodine containing contrast media favoured for its isomolar properties. With normal renal function, 97% is excreted unchanged in urine within 24 h. [2,3] The risk for sialadenitis is directly related to serum iodide levels (> 10 mg/100 mL) and inversely related to normal renal function.2 ,3 W ith increasing renal dysfunction, the elimination halflife is prolonged. Our patient was at an increased risk because of her end-stage renal failure. The delay in dialysis for the first 24 h after fistulogram might have contributed further in pathogenesis. There are also a few published case reports of pancreatic mumps and transient thyroid dysfunction; whose etiology thought to be similar to iodide induced sialadenitis. Our patient did not have any of these symptoms including thyroid swelling, abdominal pain, back pain and nausea and vomiting. Amylase, lipase, TSH and free T4, T3 were essentially normal. Role of steroids in the management of contrast induced sialadenitis has been found to be controversial in published case reports. Usual prophylactic regimens for iodide allergy were also ineffective in a previous case report proving it more idiosyncratic reaction than hypersensitivity.[4] The reaction is probably a class effect, because substituting one form of lowosmolar nonionic contrast media for another did not prevent recurrences of the condition.

The way to prevent recurrence of iodide mumps is to avoid using intravenous iodinated contrast in those patients again or urgent dialysis within 24 hours if the use of iodinated dye is absolutely essential.[5] There is absolutely no role of antibiotic in the management of iodide mumps as the elevated WBC count is always reactive in nature.

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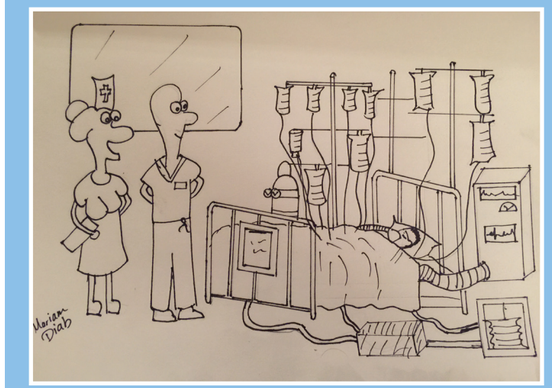
CAPTION IT

A prize will be awarded to the resident who can provide the most original and amusing caption for this cartoon created by our very own Mariam Diab. Responses should be submitted to researchjournal@stvincentcharity.com. Submissions will be accepted until October 1st 2015.



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