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Case Report

Arachnoiditis ossificans with syringomyelia: a unique case of myelopathy

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ABSTRACT

Arachnoiditis ossificans (AO) is a rare disorder characterized by calcification of arachnoid membranes first described by Kaufman and Dunsmore in 1971. It is a very rare cause of spinal canal stenosis leading to neurological compromise presenting with progressive lower extremity myelopathy. It has been described to be a sequela of various conditions previous intradural surgery, myelograms, vascular malformations and adhesive arachnoiditis. Associated conditions may include syringomyelia. The imaging findings on MRI may be confusion. Preferred diagnostic method is non contrast computed tomography (CT). Surgical intervention is still controversial and can include decompression. The authors report the case of 48 years female presenting with gradually progressing paraparesis. Magnetic resonance imaging of the spine revealed a spinal cord syrinx but with an extramedullary intradural hypo-intensity. A computed tomography scan clearly demonstrated the abnormality and its extent. We present a unique case of syringomyelia resulting from spinal arachnoiditis ossificans and review the relevant literature. This case reports a unique presentation of arachnoiditis ossificans with syringomyelia in which etiology is not clear. We also highlight the difficulty is diagnosis on MRI and need of non-contrast CT.

Keywords: Arachnoiditis, Arachnoiditis ossificans, Bony metaplasia, Syringomyelia

INTRODUCTION

Arachnoiditis ossificans is a rare clinical entity in which arachnoid cells undergo bony metaplasia secondary to end-stage adhesive arachnoidtis.[1] Spectrum of calcified meningeal plaques exist ranging from small focal calcifications to thick, intrathecal calcifications seen in arachnoiditis ossificans. Small plaques are a result of chronic degenerative processes and present little clinical concern because they are almost always asymptomatic. On the other hand, large plaques progressively compress the neural elements to produce severe neurological sequelae.[2-4] AO is a very rare entity, with limited number of reports in literature. Given these limitations there is no consensus regarding its therapeutic management guidelines. Some authors have advocated improvement in neurologic symptoms post laminectomy and after removing calcified plaques, while others don't support supportive line of treatment. We present a patient that illustrates the challenges in the diagnosis and management of AO.

Clinical details

A 48 years female presented with neck pain and gradually progressive weakness in bilateral lower limbs over a period of 2 years. Physical examination revealed mild wasting of her quadriceps muscles and the gastrocnemius muscles. Muscle motor testing revealed 3/5 paraparesis. A sensory deficit could be ascertained at

the T-8 level. Joint position and vibration senses were diminished below the knees. The patellar and Achilles tendon reflexes were bilaterally hyperactive. Past history revealed she was treated for pulmonary tuberculosis 22 years back. She again presented in Oct 2018 with complaints of cough with expectoration, breathlessness and fever. Sputum culture was negative for tuberculosis. This time she responded well to antibiotics and conservative management. She is a known case of DM-2 and HTN.

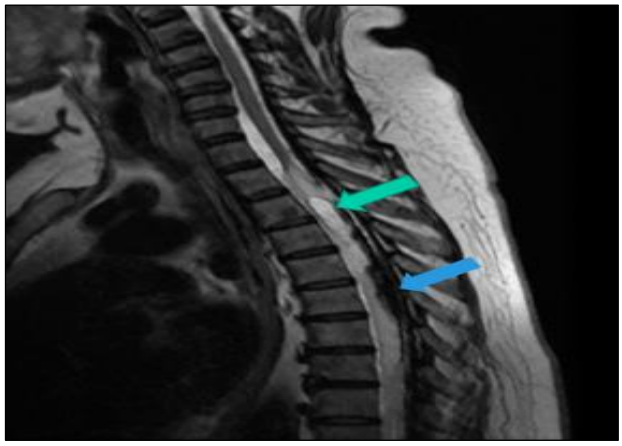

Figure 1: T2 weighted sagittal MRI of dorsal spine-shows a hypointense extramedullary intradural lesion over dorsal aspect in mid dorsal cord region leading to anterior displacement and compression of cord with cord edema (blue arrow). Also, syrinx was noted within the cord adjacent to it (green arrow).

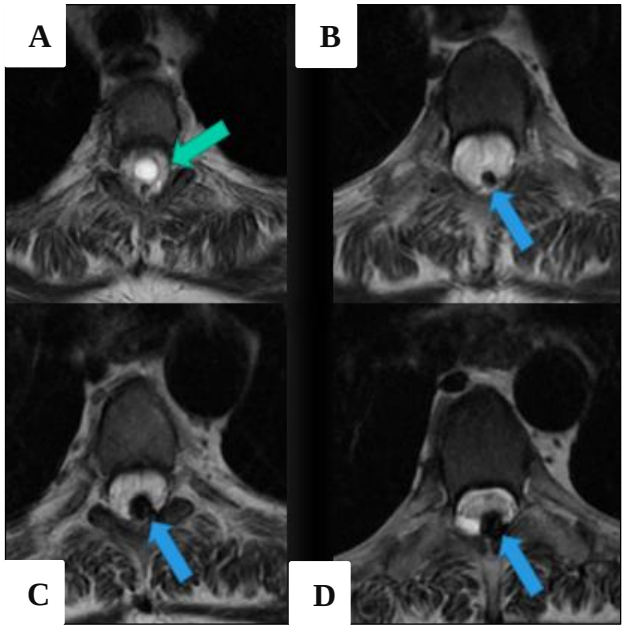

Figure 2: T2 weighted axial MRI (cranial to caudal) of dorsal spine -shows a syrinx cranially (green arrow) (A) and a T2 hypointense extramedullary intradural lesion over dorsal aspect leading to anterior displacement and compression of cord with cord edema (blue arrow) (B, C and D).

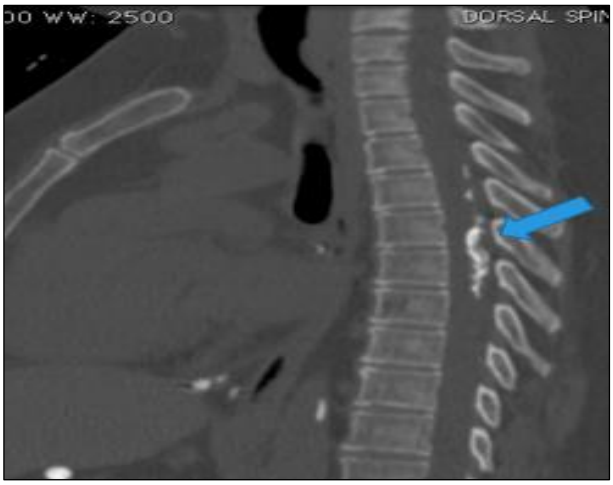

Figure 3: Sagittal CT of dorsal spine-shows a long segment ossification of arachnoid membranes in mid dorsal cord (blue arrow).

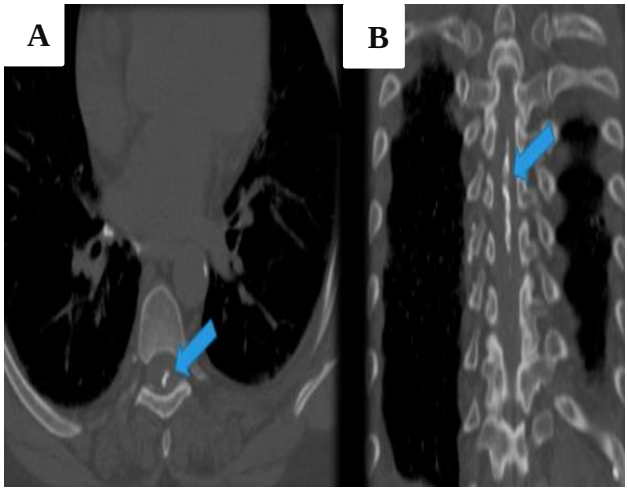

Figure 4: Axial (4a) and coronal (4b) CT of dorsal spine - shows ossification of arachnoid membranes in mid dorsal cord.

MRI was done for further evaluation which showed a T1/T2 hypointense extramedullary intradural lesion over dorsal aspect in mid dorsal cord region leading to anterior displacement and compression of cord with cord edema. No enhancement was seen on post-contrast study. Syrinx was noted within the cord opposite to this lesion (Figure1 and 2). Also signs of arachnoiditis were seen in lower cervical cord in form of focally expanded anterior sub-arachnoid space with scalloping of posterior part of vertebral bodies. MRI appearance suggested calcification/ossification or hemosiderin deposition from prior trauma. Subsequently, a CT scan of cervicodorsal spine was obtained which showed a long segment ossification of arachnoid membranes in mid dorsal cord compatible with arachnoiditis ossificans (Figure 3 and 4). Also, multiple large calcified mediastinal, hilar and abdominal lymph nodes were seen likely sequele of old tubercular infection.

DISCUSSION

Arachnoiditis ossificans (AO) is a unique and uncommon phenomenon characterized by intrathecal bony metaplasia of the arachnoid membrane.[4]

A number of insults have been associated with arachnoiditis ossificans including prior trauma, spinal surgery, subarachnoid hemorrhage, pant opaque myelography, spinal anesthesia. It has been postulated that mature bone cells arise from multipotent arachnoid cells secondary to insults mentioned above.[4,5] However; etiology in our case is not entirely clear. Though she has a history of pulmonary and abdominal tuberculosis twenty-two years back, there were no spinal complaints back then. Also, history of lumbar puncture is present few years back so a possibility of sub-arachnoid hemorrhage could be considered.

Pathogenesis of syringomyelia in the setting of AO has been hypothesized to result from vessel ischemia, CSF flow alterations, or even as a coexisting incidental finding. Scarring of the arachnoid membranes causes changes in the vascular supply producing areas of ischemia with subsequent cavitation of the spinal cord. Progression of the syrinx cavity results from altered CSF flow dynamics and subsequent spino-spinal pressure dissociation and expansion of syrinx cavities.[6]

Clinically patient may present with low back pain, radicular or nonradicular leg pain, paraparesis and bladder and bowel dysfunction.

On imaging, most common location is thoracic spine, correlating with highest concentration of arachnoidid cells, followed by the lumbar spine.[7] Bone CT shows calcific densities within clumped nerve roots. Also, calcifications may surround conus medullaris, cauda equine. On MRI, T1WI shows areas of ossification are of variable signal, i.e., hypointense, isointense, or hyperintense. On T2WI clumped nerve roots forming cords and mass(es) with secondary adhesions and arachnoid cysts are seen. Linear or globular hypo intensity if calcifications are present. Larger areas of ossification can occasionally be hyperintense on T2WI which may exert mass effect on cord, conus and cauda equine. On T1WI C+ minimal or no nerve root enhancement is seen.[1,4-7]

AO was divided by Domenicucci M et al, in 2004 into three types based on computed tomography (CT) and magnetic resonance (MR) findings-type I ossifications are semicircular in appearance, type II is circumferential, and type III are a honeycomb like ossifications, which affect the cauda equina.[8]

The management of arachnoiditis ossificans remains a dilemma between conservative and surgical treatment.[9] Patients with mild symptoms should be followed closely. In patients with severe symptoms, surgery is often performed with the target of decompression of the neural elements. Around 50% of the case with surgical intervention showed improvement if one goes by the literature. Very few cases of arachnoiditis ossifications with syringomyelia have been reported till date, therefore the standard treatment/intervention has not been established. Laminectomy and decompression with or without syrinx drainage and shunting remain the major surgical techniques. Different views exist regarding the need for drainage and shunting of the syrinx.[4,6,9,10]

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