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Review

Arachnoiditis ossificans of the thoracic spine

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ABSTRACT

Arachnoiditis ossificans is a rare disorder characterized by the development of calcifications of the arachnoid membrane of the thoracic and lumbar spines. It is an extremely rare cause of spinal canal stenosis and consequent neurological compromise, and its origins and optimal management remain unclear. We review of the literature that illustrates the challenges of diagnosis and treatment of arachnoiditis ossificans. A patient with arachnoiditis ossificans is discussed to illustrate the presentation, treatment, and prognosis of the disease.

1. Introduction

Calcified leptomeningeal plaques occur on a spectrum of severity. On one end of the spectrum exist small calcifications, which can be found in up to 78% of autopsies [1]. These plaques appear to be the result of chronic degenerative processes and present little clinical concern because they are almost always asymptomatic. On the other end of the spectrum are the thick, intrathecal calcifications seen in arachnoiditis ossificans (AO). These plaques have the capacity to progressively compress the neural elements to produce severe neurological sequelae. AO is a rare entity, having been described in the literature fewer than 70 times to our knowledge. Given the infrequency with which this disease occurs, the clinical community has yet to reach a consensus about how AO should be approached therapeutically. Some advocate for an aggressive surgical intervention, while others are less eager to take patients with AO to the operating room [2,3]. We present a patient that illustrates the challenges in the diagnosis and management of AO.

2. Illustrative patient

A right-handed 48-year old woman was referred to our neurosurgery clinic for evaluation of left-sided weakness that had been progressing over the previous two decades. Twenty-three years prior the patient was diagnosed with a giant basilar artery aneurysm with obstructive hydrocephalus. This was treated with suboccipital craniotomy and C1–2 laminectomies for bilateral vertebral artery ligation at another institution. She experienced an intraoperative stroke that produced a visual field deficit, diminished gag reflex, and left-sided weakness. Since that time, the patient underwent several procedures including syringosubarachnoid shunt for a cervical spine syrinx, and fourth ventricular shunting. Despite these interventions, the patient continued to experience pain and progressive weakness of her left extremities. Over the past 3 years she experienced worsening gait dysfunction and lost her ability to ambulate unsupported.

At her most recent clinical presentation, she had full strength on the right side. On the left, her strength was 5/5 in her quadriceps and plantar flexion and between 3/5 and 4/5 her deltoid, triceps, bicep, grip, iliopsoas, hamstrings, and dorsiflexion. She had mild spasticity in her left upper and lower extremities, with normal tone on the right. In addition, she had an up-going Babinski reflex and a positive Hoffman sign in her left extremities. Her Achilles, patellar, biceps, triceps, and brachoradialis reflexes were 2+ bilaterally. She walked unsteadily using a cane with significant circumduction of the left lower extremity.

An MRI was performed, but failed to demonstrate a cause for the patient's progressive neurologic decline. Therefore, a CT myelogram was performed to evaluate her cervical spine. Despite multiple positioning attempts during the myelogram, contrast would not extend superior to the T7–8 interspace level (Fig. 1A). A CT scan of the cervical and thoracic spine was ordered to follow the myelogram to further characterize the rostral extension of the contrast. The thoracic CT myelogram demonstrated asymmetry of the contrast in the inferior-thoracic levels, and only a small amount of contrast could be appreciated in the posterior thecal sac in the mid-thoracic levels (Fig. 1B). The cervical CT scan was relatively unremarkable. After thorough discussion with the patient, she was taken to the operating room to explore the etiology of her significant myelographic block.

A T3–6 laminoplasty was performed with intraoperative monitoring of sensory and motor evoked potentials of the lower extremities. When the dura was opened along the dorsal midline, a thick

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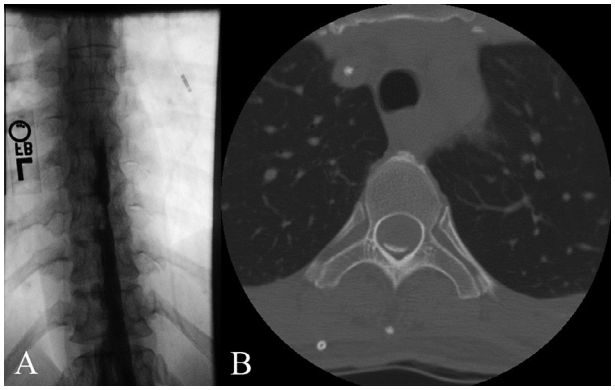

Fig. 1. Radiographic imaging of the patient before her first operation. (A) A plain film myelogram demonstrates that no contrast is able to pass rostral to the T7–8 interspace despite multiple positioning attempts. (B) An axial CT scan of T4 taken shortly after the image in (A) demonstrates a hyperdense stripe in the posterior thecal sac which could be consistent with either myelogram contrast or calcification.

hard shell of calcium consistent with AO was encountered. The superior lip of the shell, which was at the T3 level, was gently explored. Using the operating microscope and microdissectors, this superior margin of the calcium plaque was gently freed from the underlying spinal cord. The excision of the plaque was continued caudally with the 2 mm Kerrison punch until the shell could no longer be visualized. The plaque was approximately 3 mm thick, and it extended the entire length of the exposure from T3 to T6. The spinal cord beneath the shell was compressed. Once the visible plaque had been completely removed, ultrasound was used to evaluate the thoracic syrinx, but scattered arachnoid calcifications limited this study. A small midline myelotomy was made to evaluate the syrinx, but the cavity was found to be small and insufficient to drain. In the immediate postoperative period the patient had decreased strength in her right lower extremity and decreased sensation in her bilateral lower extremities. This slowly improved, and at her 3 month follow-up, the patient felt that her strength in both lower extremities was gradually returning.

Follow-up MRI at 4 months showed a significantly enlarged thoracic cord with increased T2 signal compared to preoperative studies (Fig. 2A). A CT scan demonstrated that the plaques, which now were seen to extend from C7 to T11, involved much more of the spinal cord than originally thought (Fig. 2B). Given the slow progress in her motor recovery and the more widespread findings on imaging, the patient elected to undergo another, more extensive operation to resect the compressing ossification. A laminoplasty from T1 to T11 was performed, and the inferior edge of the plaque was identified at the T11 level. The plaque was carefully removed by following the plane between the calcification and the spinal cord (Fig. 3; Supp. Video1). In segments where the plaque encompassed much of the circumference of the spinal cord, a Kerrison punch was used to dissect the midline portion of the calcification, thereby dividing the plaque into two halves that could be carefully removed. Upon dissecting the plaque away, the thoracic cord became much more pulsatile. The resected plaque measured more than 6 inches in length (Fig. 3). After 4 weeks of physical therapy, the patient returned to clinic and described significant progress in regaining strength and function. Four months after her second decompressive surgery, the patient had an intrathecal baclofen pump implanted for residual spasticity.

3. Review of the literature

The pathogenesis of AO has not been fully elucidated. It is considered to be the endpoint of an inflammatory process starting

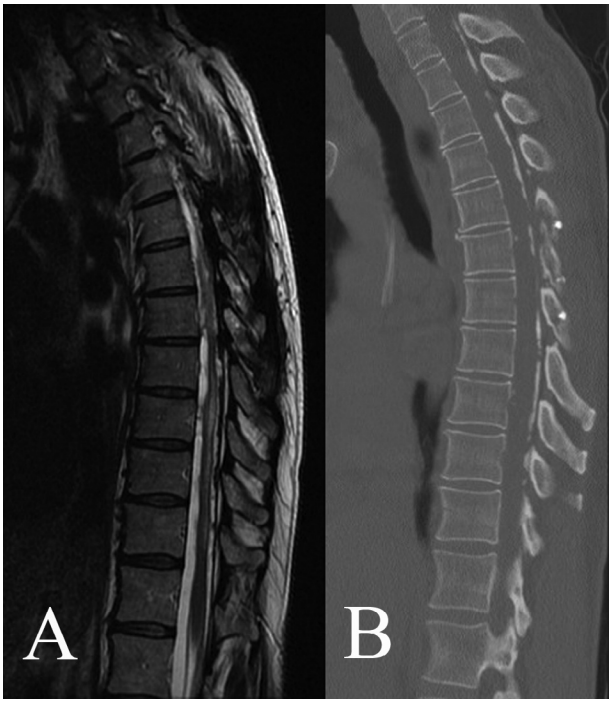

Fig. 2. Imaging of the patient 4 months after the first operation. (A) T2-weighted sagittal MRI demonstrates increased T2 hyperintense signal changes within the thoracic cord consistent with compressive myelopathy. (B) Sagittal CT scan reveals that the arachnoid calcification extends in the dorsal spinal canal from T1–11.

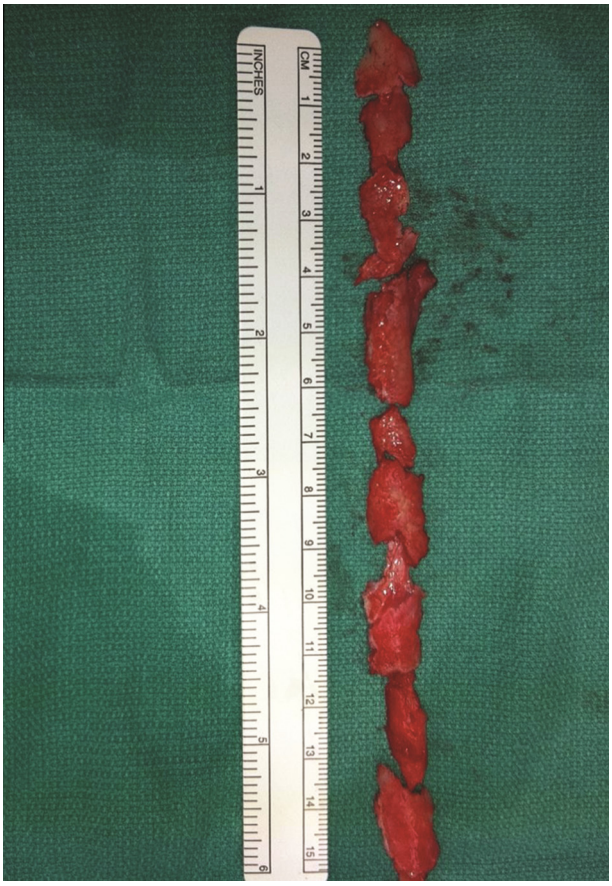

Fig. 3. Specimen from the patient's second operation. The entirety of the resected arachnoid calcification, which extends beyond 6 inches in length. This figure is available in colour at www.sciencedirect.com.