Arachnoid ossificans containing metaplastic hematopoietic marrow resulting in diffuse thoracic intrathecal cysts and severe myelopathy

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Abstract

Objective To present a rare case of multiple compressive thoracic intradural cysts with pathologic arachnoid ossification, review the literature and present the surgical options.

Summary of background data Few reports have identified the existence of arachnoid calcifications and intrathecal cysts causing progressive myelopathy. The literature regarding each of these pathologies is limited to case reports. Their clinical significance is not well studied, although known to cause neurologic sequelae.

Methods An 81-year-old female clinically presents with rapidly progressive myelopathy. Pre-operative magnetic resonance imaging identified multiple compressive thoracic intrathecal cysts. Surgical exploration and decompression of these cysts identified calcified plaques within the arachnoid. Histopathologic examination revealed fibrocalcific tissue undergoing ossification with bone marrow elements.

Results Due to progressive myelopathy, the thoracic cysts were decompressed and calcified plaques were excised, once identified intra-operatively.

Conclusions On last examination, the patient's neurologic status had not improved, but had stabilized. The rate of neurologic improvement from excision and decompression

is variable, but it may still be warranted in the face of progressive neurologic deficits.

Keywords Arachnoid ossification · Intrathecal cysts · Myelopathy · Osseous metaplasia

Introduction

A literature search of leptomeningeal calcified plaques or “arachnoid ossificans” renders a modicum of case reports. Arachnoid calcifications have been found in autopsy studies with a reported incidence of 6.5–76% [1–4], though few reports have identified the co-existence of arachnoid calcifications with intrathecal cysts [5, 6]. Even fewer reports associate these plaques with compressive intrathecal cysts causing neurologic compromise. Although the exact pathogenesis of these calcified plaques are unknown, they have been theorized to be a result of degeneration, prior surgery, infection, congenital conditions, inflammatory or vascular anomalies, and trauma [1–3, 5, 7–13]. Histologically, the arachnoid undergoes chronic metaplastic changes leading to adherent proliferative calcified plaques. The significance of this metaplasia is not known.

Despite attempts to identify the etiology, overall pathogenesis, clinical course, and outcomes, there is very little known about arachnoid ossificans. The few reports that have been published demonstrate poorer outcome in patients with spinal cord compression. Surgical decompression of diffuse arachnoid ossification has led to mixed results and does not universally reverse the clinical course [1, 3, 7–10].

In this case study, we present a patient who presented with progressive myelopathy from arachnoid ossificans and diffuse intrathecal cysts. The present report not only adds

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to the paucity of literature, but also describes the extremely rare finding of cancellous bone within the arachnoid calcified plaques [14].

Case report

An 81-year-old female presented with a progressive gait imbalance and frequent falls. Her condition became quite disabling over the 3 months prior to presentation. On examination, the patient was myelopathic. She was unable to perform tandem gait and required maximal assistance with ambulation. She reported numbness in the anterior aspect of her right thigh and leg. She demonstrated weakness in her right quadriceps and brisk lower extremities reflex with sustained clonus, bilaterally.

Initial evaluation included full length standing radiographs (Fig. 1), and an MRI of her cervical spine, which revealed upper thoracic intrathecal cysts. A thoracic MRI

was promptly obtained (Fig. 2). Pre-operative MRI demonstrated loculated cystic lesions within the dorsal aspect of the thoracic canal extending from T2 to T10. The cysts appeared to compress the spinal cord throughout these levels with the most significant compression at T10 (Fig. 3). Due to progressive neurologic decline, the decision was made to surgically decompress the thoracic spine and excise the cystic lesions.

Spinal cord decompression was achieved with en bloc laminectomy at the involved levels and stabilized with pedicle screw fixation. Due to the amount of bony resection and facet removal during the en bloc laminectomy, it was determined that pedicle screw fixation and fusion were necessary to minimize risk of post-operative instability and iatrogenic kyphosis. An extensive midline durotomy was then performed. The microscope was brought in for microsurgical dissection and the dura was tacked up. Extensive calcifications and adhesions were contiguous with the arachnoid; although they were not adherent to the spinal cord. The lesions clearly deformed the spinal cord, causing severe spinal cord compression and displacement (Fig. 4). Utilizing very meticulous microsurgical dissection, a plane was slowly developed to separate the calcifications from the spinal cord. The lesions also adhered to virtually every exiting root in the thoracic spine and some of the lesions

Fig. 1 Pre-operative full length standing 36". **a** AP, **b** lateral films

Fig. 2 Pre-operative mid-sagittal thoracic MRI identifying loculated cystic lesions (arrow) within the dorsal aspect of the thoracic canal extending from T2 to T10

Fig. 3 Pre-operative MRI with axial view of T10. The dorsal intrathecal cyst is compressing the spinal cord most significant at this level (*arrow*)

were unable to be fully dissected for fear of causing neurological injury. No changes in neurophysiologic monitoring were appreciated during any portion of the surgery. Once the calcifications were fully resected, the spinal cord returned to a normal shape and the dura was closed in a continuous fashion. A posterior fusion with instrumentation was performed due to the extensive resection of the posterior elements and partial resection of facet joints.

Grossly, multiple arachnoid calcifications were sent to pathology for immediate evaluation. Histopathologic analysis revealed lamellar bone containing mature adipose tissue and hematopoietic bone marrow elements, including megakaryocytes (Fig. 5).

Pathology gross and microscopic methods and analysis

The specimens were placed in formalin fixative for paraffin embedding following decalcification. Histologic sections showed fibrocalcific tissue undergoing ossification with scattered psammomatous calcifications and rare arachnoid cells. The areas of osseous metaplasia contained hematopoietic bone marrow elements and mature adipose tissue (Fig. 5).

Post-operative course

The patient was monitored in the intensive care unit immediately following surgery. A chest radiograph demonstrates the gross view of the operative fixation (Fig. 6).

Fig. 4 Intra-operative examination identifying extensive calcifications (*arrows*) and adhesions present within the arachnoid causing severe spinal cord compression and displacement

Fig. 5 Histopathologic specimen revealed lamellar bone containing mature adipose tissue and hematopoietic bone marrow elements, including megakaryocyte (*arrow*)

A CT evaluation was obtained to ensure proper placement of instrumentation (Fig. 7). The patient's neurologic status stabilized and she was cleared to work with physical and

occupational therapy. A post-operative MRI was later obtained, but not included due to the significant artifact.

On a 1-month post-operative evaluation, she continued to have profound weakness in her right quadriceps graded 1/5. The remainder of her right lower extremity on manual muscles strength testing demonstrated 3/5 in her tibialis anterior strength, 4/5 in both her extensor hallucis longus and plantar flexion. Her left lower extremity demonstrated full strength for the exception of her quadriceps, graded as 3/5. Regarding her sensation, she had less sensation globally to bilaterally lower extremities. Clonus to bilateral lower extremities remained. Overall, her strength had diminished from her pre-operative status and her gait was significantly impaired due to both her myelopathy and weakness.

Fig. 6 Post-operative chest X-ray performed in the intensive care unit, demonstrating pedicle screw instrumentation

Discussion

We present a patient with progressive myelopathy and multiple compressive thoracic intradural cysts. She was taken to the operating room for posterior decompression and fusion. Pathological analysis revealed arachnoid ossificans with cancellous bone within the arachnoid calcified plaques. Osseous metaplasia with subsequent bone marrow formation has been described and usually occurs around areas containing dystrophic calcified tissue. The conversion of fibrocellular interstitial tissue to adipose or hematopoietic marrow is seen in the late development of osseous metaplasia [14]. Many factors may predispose arachnoid cells to undergo metaplasia. Whether due to vascular abnormalities or repetitive insult, the multipotential cells are exposed to a hypoxic environment and undergo bony metaplasia. The proliferation of osteoblasts leads to the development of ossification [6, 8, 10, 15].

When arachnoid calcifications become ossified (arachnoid ossifications), Wijdicks [16] reported that patients might present with a constellation of clinical symptoms including radicular pain, spastic paraparesis or cauda equina syndrome. Based on autopsy reports, though, the gross majority of patients with arachnoid calcifications are asymptomatic. According to the report by Slavin et al. [17], arachnoid calcifications can present in one of three ways. It may present in asymptomatic patients at the time of surgery or autopsy, as isolated calcified plaques or as diffuse ossification of the arachnoid membrane at several segments of the spinal cord, which was present in the current case. Removal of these plaques has reported to improve the clinical neurologic deterioration in 43% of operated patients according to one literature review [3, 6, 13, 18, 19]. According to a review by Whittle et al. [1, 3], however, these benefits have been minimal in 42% of reported cases.

Fig. 7 Axial images of a post-operative CT at **a** T2–T3, **b** T10–T11 (note the drain in place)

Excision of plaques and diffuse arachnoid plaques failed to demonstrate long-term improvement in reports by Sakai et al. [5] and Papavlosopoulos et al. [6]. Additional reports of cyst excision without arachnoid ossification have had favorable results [20–22]. Unfortunately, there is a paucity of reports of arachnoid ossification with diffuse arachnoid cysts to demonstrate consist surgical outcomes.

There is currently no standard of care regarding treatment of diffuse intradural cysts and leptomeningeal calcifications, especially since their coetaneousness has only been reported twice previously. Progressive neurologic deterioration warrants surgical treatment [8]. Additional methods of management for intradural cysts include percutaneous drainage or shunting the fluid to the peritoneum or pleura [5, 23–25]. Excision of calcifications and neurolysis is technically difficult and surgical outcomes are variable; therefore, surgical intervention is reserved for neurologically compromised patients. Asymptomatic or mildly symptomatic patients should be treated with close observation.

To our knowledge, there are no reports on the co-existence of metaplastic bone marrow in arachnoid ossificans with diffuse arachnoid cysts. The severity of compression due to both arachnoid calcifications and spinal arachnoid cysts can lead to myelopathy, radiculopathy, or cauda equina syndrome. Although the rate of neurologic improvement from excision and decompression is variable, it may still be warranted in the face of progressive neurologic deficits.

Conflict of interest None of the authors has any potential conflict of interest.

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