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Case Report

Scalpellum occulta: A rare case of dorsal spinal arachnoid web without the scalpel sign ☆,☆☆

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ABSTRACT

Spinal arachnoid web is a rare condition characterized by extramedullary bands of arachnoid tissue at the level of the dorsal thoracic spinal cord that may lead to progressive, permanent neurological deficits. To date, this condition has been radiographically characterized by a scalpel sign, which has been pathognomonic in all reported cases of spinal arachnoid webs. In this case, we report the first known patient with confirmed spinal arachnoid web without radiographic evidence of the scalpel sign. In reporting our finding, we encourage a higher clinical suspicion for spinal arachnoid web in patients presenting with progressive thoracic myelopathy following trauma, and radiographic evidence of ventrally displaced spinal cord and turbulent cerebrospinal fluid flow, even in the absence of a scalpel sign.

Introduction

A spinal arachnoid web (SAW) is an extremely rare condition marked by thickened extramedullary bands of arachnoid tissue in the dorsal thoracic spinal cord [1]. Though isolated cases of SAWs have previously been attributed to spinal cord trauma, infection, or surgery, no definitive etiology or associated risk factors have been identified [2,3]. The clinical presentation of SAWs varies across patients. While some individuals

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may present asymptomatically, others may present with paraplegia or paraparesis, hypoesthesia, and bladder/bowel dysfunction over the course of months to years [2,4]. Although the visualization of arachnoid lesions on imaging remains challenging due to resolution constraints, a pathognomonic feature present in SAWs is the scalpel sign, which is a characteristic indentation on the dorsal surface of the thoracic spinal cord seen on magnetic resonance imaging (MRI) [2]. In this case, we report the first known patient with SAWs without radiographic evidence of the scalpel sign.

Case report

A 19-year-old female with no significant past medical history presented to our institution for evaluation of bilateral leg weakness and paresthesias for the past 8 months following a mechanical fall on her lower back while dancing. Her Oswestry Disability Index score was 68%, with neurological exam revealing bilateral 3/5 lower extremity (LE) strength, LE apraxia, paresthesias below the T7 dermatome, and symmetric LE hyperreflexia. Initial MRI of the thoracic spine demonstrated multiple flow voids at the midthoracic level, most prominent at T8, concerning for dural arteriovenous fistula, which was ruled-out with spinal angiogram. Subsequent imaging including 2 spinal MRIs and a computed tomography (CT) myelogram were also read negative for pathology.

During our retrospective evaluation, her latest spinal MRI demonstrated a ventrally located cord with thoracic stenosis and turbulent adjacent cerebrospinal fluid (CSF) flow, although without evidence of extrinsic cord compression, neural foraminal narrowing, syrinx, or scalpel sign (Figs. 1A-E). On 2 prior spinal MRIs, flow abnormalities were again retrospectively seen at the affected thoracic region and as serpiginous CSF filling defects on CT myelogram, all

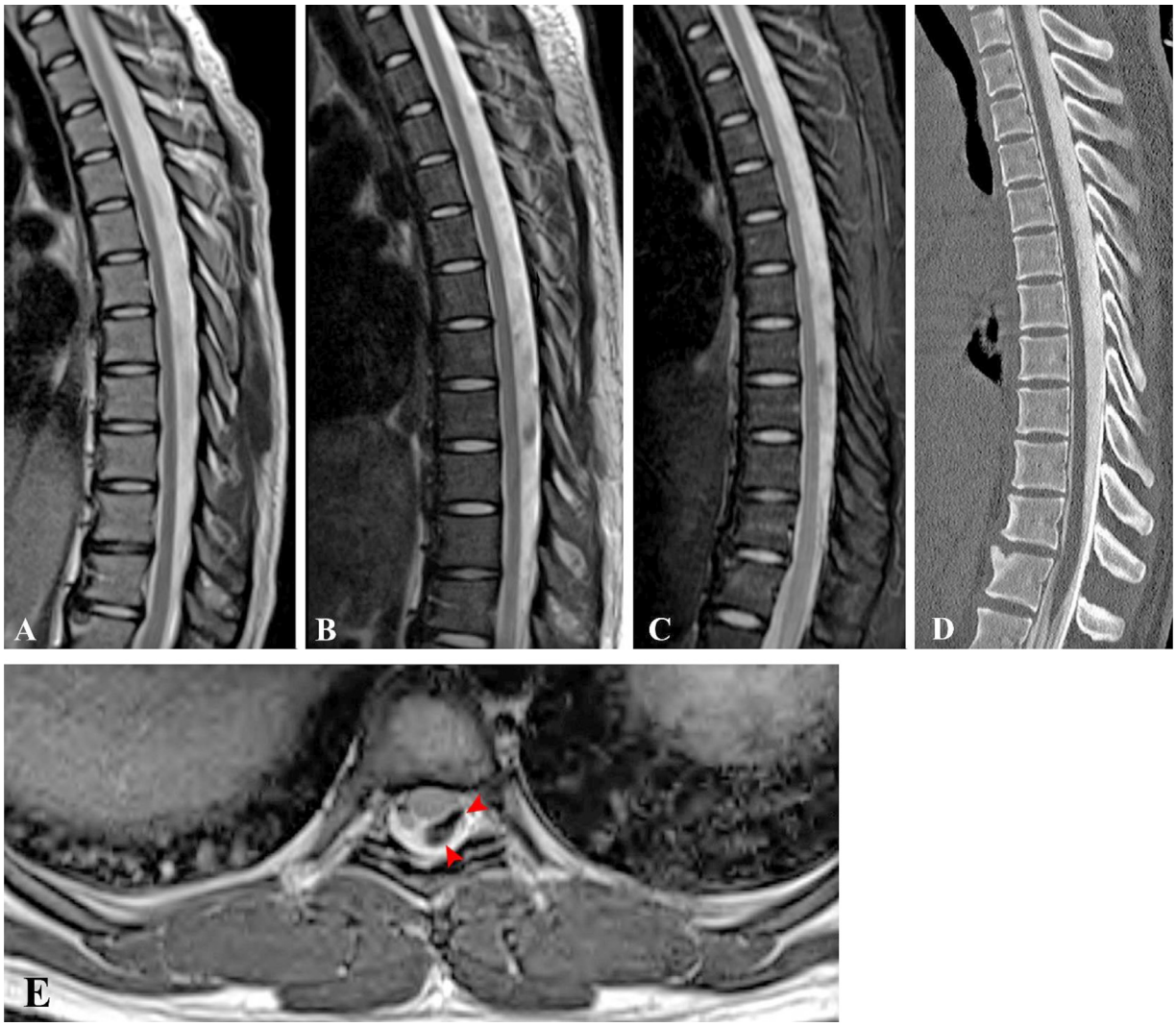

Fig. 1 – Evidence of CSF flow abnormalities are demonstrated on sagittal T2-weighted MRI reformats of the thoracic spine at the following time-points: (A) initial injury, (B) 3-month postinjury, and (C) 4-month postinjury. (D) CT myelogram of the thoracic spine demonstrates serpiginous CSF filling defects. (E) Axial T2-weighted MRI of the thoracic spine showing CSF flow abnormalities (red arrows). CSF, cerebrospinal fluid; CT, computed tomography, MRI, magnetic resonance imaging.

of which were deemed artifactual at the time of the initial interpretation (Figs. 1A-E). In the setting of her long-term symptoms and radiographic evidence of spinal cord stenosis with surrounding flow abnormalities, the patient underwent an exploratory posterior T7-9 decompression demonstrating arachnoid webbing at the T8 level, which were subsequently resected to ensure free CSF flow. The patient's postoperative course was unremarkable, with notable improvement and an Oswestry Disability Index score of 13% at a 6-month follow-up visit.

Discussion

In this case, we describe the first documented patient with symptomatic dorsal SAW without a radiographic scalpel sign on MRI. Prior to this report, all documented cases of SAW demonstrated evidence of the scalpel sign, which was considered pathognomonic of an intradural, extramedullary spinal pathology [5]. The sensitivity and specificity of the scalpel sign for diagnosis of SAW however remain undetermined due to the rarity of this condition. The scalpel sign is thought to arise from anterior displacement of the cord due to a widened posterior subarachnoid space, thus resembling the profile view of a surgical scalpel on sagittal MRI [2]. Our suspicion for a radiographically silent spinal mass was influenced by a ventrally positioned thoracic spinal cord and altered CSF flow dynamics localizing to the same thoracic region on multiple spinal MRIs. As demonstrated in this case, radiographic features of a suspected spinal mass should increase clinical suspicion for SAW even without a scalpel sign.

Differential diagnosis of the scalpel sign includes spinal arachnoid cysts and ventral spinal cord herniation [6,7]. Some distinguishing features of arachnoid cysts on imaging are a defined wall and a protracted filling pattern during myelography compared to the turbulent flow seen in SAW [2,8]. SAWs are also more commonly associated with syringomyelia, with the syrinx thought to be formed from alteration of the intramedullary pulse pressure, leading to formation of a pressure gradient from the center of the cord relative to the subarachnoid space [9,10]. In comparison, ventral cord herniation on imaging manifests as asymmetric cord protrusion through a ventral dural defect, producing a C-shaped cord on MRI, compared to the symmetric anterior displacement seen with dorsal SAWs [6].

The definitive treatment for SAWs is surgical management, although a more conservative approach may be deemed appropriate as well for patients presenting with less pronounced clinical manifestations [5]. The most common surgical intervention for patients with dorsal SAWs is laminectomy with excision of the intradural arachnoid web; however syringopleural shunt or stent placement may also be considered if the web is located circumferentially or ventrally [5,11,12]. Patients generally experience excellent outcomes, with up to 91% of patients demonstrating improved neurological status postoperatively [5]. While no specific guidelines exist, asymptomatic patients with an incidental discovery of SAW may choose to undergo close clinical observation [5,9].

Conclusion

SAW is an extremely rare condition with a poor prognosis that can lead to severe neurological impairment if left untreated. Although SAW is strongly associated with the scalpel sign on sagittal MRI, its absence does not preclude the diagnosis as demonstrated in our case report, underscoring the importance of integrating clinical suspicion and additional radiographic findings in the diagnosis of nonpathognomonic SAW. In a patient presenting with progressive thoracic myelopathy, radiographic evidence of a ventrally displaced spinal cord and turbulent CSF on multiple separate time-points should prompt consideration of SAW, even in the absence of a scalpel sign.

Patient consent

Informed written consent was obtained from the patient for the publication of this case report.

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