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Thoracic spinal arachnoid web and syringomyelia with rostral expansion to the first cervical spinal cord level: Case report



Christian Nieves-Ríos^{a,*}, Josué G. Layuno-Matos^a, Gerardo Olivella^b, Norman Ramírez^c, Carlos Weber-Seda^d, Jorge Lastra-Power^d

^a Department of Surgery, Ponce Health Sciences University, Ponce 00716, Puerto Rico

^b Department of Orthopaedic Surgery, University of Puerto Rico Medical Sciences Campus, San Juan 00921, Puerto Rico

^c Department of Pediatric Orthopaedic Surgery, Mayaguez Medical Center, Mayaguez 00681, Puerto Rico

^d Department of Neuroscience, Manati Medical Center, Manati 00674, Puerto Rico

ARTICLE INFO	A B S T R A C T
Keywords: Arachnoid web Syringomyelia Scalpel sign Cervical spine Case report	Introduction and importance: A spinal arachnoid web is a rare pathology that has been associated with the development of syringomyelia. Syrinx expansion can occur, which can result in the development of new symptoms. In the current literature, the farthest rostral expansion of the associated syrinx has been to the C3 spinal level. <i>Case presentation:</i> We present a 49-year-old Hispanic male with a thoracic spinal arachnoid web and an associated syrinx spanning from C1 to T7 spinal level. The patient developed upper extremities radicular symptoms that worsened over time. He underwent surgical management with T6–8 laminectomy and excision of the web. Postoperative follow-up evaluations demonstrated progressive clinical improvement with eventual resolution of symptoms and syringomyelia. <i>Clinical discussion:</i> Syringomyelia secondary to a thoracic spinal arachnoid web is a progressive disease that can expand rostrally to the C1 spinal level. The clinical presentation usually involves the lower extremities. However, if the upper thoracic or cervical spinal cord is involved, patients can also present symptoms in the upper extremities. Management usually involves surgical excision of the web in order to decompress the subarachnoid space and restore the normal cerebrospinal fluid (CSF) flow. <i>Conclusion:</i> Our case suggests that syringomyelia secondary to a spinal arachnoid web may continuously expand with concomitant worsening of symptoms. However, surgical resection is curative with possible remission of symptoms and normalization of spinal anatomy.

1. Introduction

Spinal arachnoid webs are rare pathologic thickening of transverse bands of arachnoid tissue [1]. These bans are commonly found in the dorsal aspect of the thoracic spine and have been associated with compressive myelopathy symptoms [1,2]. Arachnoid webs were considered a variant of an arachnoid cyst, but they are a distinct entity of focal arachnopathy [1,3–5]. Given their rarity and unclear classification, little epidemiological information is currently available [6,7].

The diagnosis of arachnoid webs relies on conventional imaging techniques such as magnetic resonance imaging (MRI) and computerized tomography (CT) scans [5,8,9]. The radiological hallmark of a spinal arachnoid web is a focal dorsal indentation of the spinal cord that has been described as "the scalpel sign" [1,10]. In addition, the presence of syringomyelia is considered a supporting feature of this disease [11]. Nonetheless, these findings are often subtle and can be easily overlooked [5].

Clinically, patients can present with a wide range of symptoms, from mild weakness or numbness to paraplegia and saddle anesthesia [4]. Yet, the most common symptoms include neuropathic pain, motor weakness, and sensory loss, predominantly in the lower extremities [4,5]. Interestingly, it has been noted that patients can also manifest symptoms in the upper extremities, particularly in the presence of an associated syrinx at the cervical spinal level [11,12].

* Corresponding author at: Street 130, Parque Monterrey#2, Apt 316, Ponce 00716, Puerto Rico.

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Abbreviations: BMI, body mass index; CSF, cerebrospinal fluid; CT, computerized tomography; MRI, magnetic resonance imaging.

E-mail addresses: cnieves17@stu.psm.edu (C. Nieves-Ríos), jlayuno20@stu.psm.edu (J.G. Layuno-Matos).

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To our knowledge, only eight cases of cervical syringomyelia secondary to a thoracic spinal arachnoid web have been described [2,5,11–14]. Yet, in the current literature, the farthest rostral expansion of the associated syrinx has been to the C3 spinal level [11]. We hereby tender a case of syrinx expansion to the C1 spinal level secondary to a thoracic spinal arachnoid web. This case report has been reported in line with the Updating Consensus Surgical Case Report (SCARE) criteria [15].

2. Case presentation

A 49-year-old Hispanic male presented to our private office with a history of chronic interscapular, sternal, and right upper extremity radicular pain to the middle finger with paresthesia. The medical history was remarkable for morbid obesity (body mass index (BMI): 42.6 kg/m²). The review of systems revealed a chronic history of neck, back, and multiple joint pain. The patient also disclosed a history of numerous car accidents and falls.

On a comprehensive neurological examination, the cranial nerves' functions were within normal limits. Normal gait was observed with no motor weakness or sensory deficit. The upper and lower extremity deep tendon reflexes were normal (+1) bilaterally, and no long tract signs were identified.

The patient presented to his first visit with a one-year-old MRI that suggested the presence of a seven-centimeters long syrinx cavity from T4 to T7 (Fig. 1). Subsequently, a new MRI was ordered, which showed a T7–8 dorsal spinal indentation suggestive of an intradural arachnoid web. Moreover, a rostral expansion of the initial syrinx was identified that now spanned from T2 to T7. No changes were noted in the cervical spinal cord (Fig. 2).

Surgical treatment was recommended, but due to personal reasons, the patient decided to postpone the surgery. However, after one year, he returned with new-onset left upper extremity discomfort, weakness sensation, sensory deficit, and paresthesia.

Physical examination was remarkable for mild left upper extremity weakness (5/5 deltoid, 5/5 bicep, 4/5 tricep, 4/5 interossei, 4/5 grip)

and decreased temperature sensation. Also, a bilateral increase in knee and ankle reflexes (+3 and +2, respectively) was identified. At this visit, a third MRI showed further rostral expansion of the syrinx extending from C1 to T7, with the initial lesion still present at the T7–8 level (Fig. 3).

Due to the exacerbation of symptoms and the extension of the syrinx, the patient agreed to undergo surgical treatment by the principal investigator (spine neurosurgeon). The surgical intervention consisted of T6–8 laminectomies with intradural excision of the arachnoid web. An irregular fragment of tan whitish semitranslucent rubbery tissue measuring $1.3 \times 0.6 \times 0.1$ cm was removed. The pathological findings of the excised specimen included fibro-membranous tissue with dystrophic microcalcifications (Fig. 4).

The immediate postoperative status was remarkable for improved motor and sensory functions. However, paresthesia and left shoulder discomfort were still present, but to a lesser degree. Three months following the surgery, only residual paresthesia in his left arm was identified. At the one-year follow-up visit, the patient fully recovered with regression of all symptoms and was pleased with the results. The postoperative MRI showed complete resolution of both the syrinx and the dorsal spinal cord indentation (Fig. 5).

3. Discussion

Spinal arachnoid webs are extremely rare thickenings of the transverse arachnoid fibers that extend to the pial surface of the dorsal spinal cord [1]. Although the exact etiology of this pathology remains unknown, multiple reports have noted that arachnoid webs can develop after a traumatic event [4,5,11]. In our patient, given his history of numerous car accidents and falls, it is possible that a blunt trauma to the thoracic spine was the cause of the arachnoid web. However, in our case, we could not establish a direct relationship between a specific traumatic event and the development of the arachnoid web.

The key feature for diagnosing a spinal arachnoid web is the identification of "the scalpel sign" on imaging [1,14]. This sign describes the focal dorsal indentation in the spinal cord that gives the appearance of a



Fig. 1. (A) Sagittal T2-weighted MRI through the cervicothoracic spine demonstrates expansile signal abnormality at the T4–7 levels, manifested by intramedullary T2 hyperintense signal consistent with cord edema (bracket). There is an abrupt caliber change at the T7–8 level, with a "scalpel sign" (arrow). (B–C) Axial T2-weighted MRI through the thoracic spinal cord demonstrate expansile signal alteration in addition to a small, superimposed syrinx eccentric towards the right (asterisk). (D–E) Axial T2-weighted MRI of the mid-thoracic spine at the T7 level demonstrate anterior displacement of the thoracic spinal cord (arrowhead), implying the presence of extrinsic mass-effect upon the dorsal spinal cord. MRI = magnetic resonance imaging.



Fig. 2. (A) Sagittal T2-weighted MRI of the thoracic spine demonstrates propagation of expansile signal alteration now spanning from T2–7 (bracket), and (B) persistent abrupt caliber change of the dorsal spinal cord at the T7 level, presumably by extrinsic mass-effect from the dorsal arachnoid web. (C) Sagittal T2-weighted MRI of the cervical spine demonstrates normal caliber and signal of the cervical spinal cord, without cord edema. MRI = magnetic resonance imaging.

surgical scalpel with the blade pointing posteriorly [1,16]. Moreover, patients with a spinal arachnoid web frequently develop syringomyelia secondary to a mass effect or obstruction of normal cerebrospinal fluid (CSF) flow [1,5,11,16].

The most common clinical complaints of patients with a spinal arachnoid web are neuropathic pain, motor weakness, and sensory loss, predominantly in the lower extremities [4,5]. However, a small percentage of patients can present with symptoms in the upper extremities, particularly in the presence of an associated syrinx at the upper thoracic and cervical spinal level [4,12]. Chang et al. described a case of a 32year-old female with an arachnoid web at the T4 spinal level and an associated syrinx expanding from the upper thoracic to the lower cervical spine [12]. Clinically, the patient presented with bilateral upperextremity paresthesia, pain, and weakness [12]. Similarly, Sridharan and Heilman reported a case of a spinal arachnoid web at the T4 spinal level with a syrinx expanding from T4 to C3 [11]. In their case, the patient developed a progressive decrease in sensation of his left upper extremity [11]. Interestingly, to our knowledge, we report the first case of rostral syrinx expansion to the C1 spinal level secondary to a thoracic spinal arachnoid web.

Although challenging, identification of a spinal arachnoid web is vital because surgery is curative, with reports demonstrating neurological improvements in over 91 % of patients [4,17]. Generally, the treatment of choice consists of a laminectomy with excision of the arachnoid web, although other procedures have been attempted [4,5]. Moreover, our case suggests that symptoms can be progressive, and that clinical deterioration can occur if left untreated. Thus, special attention must be paid to the radiological and clinical findings to diagnose and treat this disease adequately.

4. Conclusion

Spinal arachnoid web is a rare pathology that is commonly found in the thoracic region of the dorsal spinal cord. Our report suggests that syringomyelia secondary to a spinal arachnoid web can progressively extend to the C1 spinal level, resulting in new neurological symptoms. In addition, our case suggests that surgical excision can be curative, which may result in the remission of symptoms and the normalization of the spinal anatomy. Additional studies are required to characterize this disease further.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

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Fig. 3. (A–B) Sagittal T2-weighted MRI demonstrating further progression of expansile signal alteration now extending proximally into the cervico-medullary junction, near the C1 level (brackets). (C–D) Sagittal T2-weighted MRI showing persistent expansile signal alteration/cord edema within the thoracic spine to the T7 level and focal dorsal indentation at the T7–8 level (arrow). (E–F) Show axial T2-weighted MRI of the upper cervical spinal cord at the C3 level demonstrating intramedullary signal alteration within the left dorsal aspect of the spinal cord (arrowhead). MRI = magnetic resonance imaging.



Fig. 4. Microphotography of the arachnoid web with hematoxylin and eosin stain $50 \times$ (A) $200 \times$ (B), showing fibromembranous tissue and dystrophic micro-calcifications (bar = 1 mm).

Ethical approval

This study is exempt from ethical approval in our institution.

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Guarantor

Dr. Christian Nieves-Ríos and Dr. Jorge Lastra-Power.

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CRediT authorship contribution statement

Nieves-Ríos: conceptualization; data curation; writing; reviewing; editing; validation. Layuno-Matos: conceptualization; data curation; writing; reviewing; editing; validation. Olivella: data curation; writing; reviewing; editing; validation. Ramírez: writing, reviewing; editing; validation, supervision. Weber-Seda: conceptualization, reviewing; editing; validation. Lastra-Power: writing, reviewing; editing;



Fig. 5. (A–B) Sagittal T2-weighted MRI showing interval resolution of cord edema within the visualized thoracic spine. (C–D) Axial T2-weighted MRI demonstrating resolution of previous extrinsic mass-effect upon the dorsal spinal cord. Posterior paraspinal incisional changes are now present at T6–8. MRI = magnetic resonance imaging.

validation, supervision.

Declaration of competing interest

The authors declare that they have no conflict of interest.

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