



Case Report

Thoracic dorsal arachnoid web with rapid onset of symptoms: A report of two cases and brief review of the literature

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ABSTRACT

Background: Thoracic arachnoid webs are a rare entity and can be challenging to diagnose with sometimes subtle radiographic findings. Arachnoid webs can cause severe cord compression with associated syrinx and resulting myelopathy, weakness, sensory loss, and bowel/bladder dysfunction. There have been a little over 60 cases total reported in the literature with only one systematic review. The cases presented here have unique features including rapid onset of symptoms, symptomatic syrinx extending into the cervical spine, and intraoperative syrinx drainage, all of which are quite rare in the current published literature for arachnoid webs.

Case Description: Here, we present two patients, a 73-year-old man and 58-year-old man presenting with different symptoms and timing of symptom progression but both with “scalpel sign” and associated syrinx present on their MRIs. Each patient underwent a laminectomy with resection of arachnoid web with complete resolution of symptoms in the first case and significant improvement in the second case. Postoperative imaging in both cases showed almost complete resolution of the syrinx.

Conclusion: Early clinical evaluation and workup followed by early surgical treatment can lead to dramatic improvement in outcomes after surgery. For patients that are symptomatic from an associated syrinx, a midline myelotomy to facilitate drainage can be considered to be done concomitantly with the arachnoid web resection.

Keywords: Arachnoid web, Dorsal, Review, Spine, Surgery, Thoracic

INTRODUCTION

Thoracic dorsal arachnoid webs of the spine are rare and often challenging to diagnose, with subtle radiographic findings. When left untreated, it can have devastating effects on a patient's neurological function, causing insidious progression of weakness and myelopathy in otherwise healthy individuals. Here, we present two case reports, with descriptions of each patient's presentation with comparisons to the published literature, to better understand this lesion. These cases have unique features including rapid symptom onset and symptomatic syrinx into the cervical spine, which are quite extraordinary.

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ILLUSTRATIVE CASES

Case 1

History and examination

Patient is a 73-year-old gentleman who presented to the VA (Veterans Affairs) neurosurgery clinic with complaints of neck pain and bilateral leg weakness that started 10 days before. He had pain that radiated into both shoulders and generalized weakness and pain in both legs. He reported trouble walking, complaining “he walked like a 95-year-old.” He denied any trauma, falls, changes in balance, problems with fine motor skills, or any bowel or bladder incontinence. Before symptom onset, he was walking 3 miles a day and was regularly active.

Prior medical history includes hypertension, Stage III chronic kidney disease, esophageal invasive adenocarcinoma status postgastrectomy and resection 1 year prior requiring no adjuvant treatment, and diabetes mellitus type 2. He had no prior spine surgeries. Laboratories were unremarkable except for elevated glucose and serum creatinine of 1.99 mg/dL. Hemoglobin A1c was 6.4%.

On examination, he was full strength in both arms and was four out of five strength in both legs. Sensation was intact to light touch throughout. He had 3+ DTRs (deep tendon reflexes) in his left arm, 2+ DTRs in the right arm, and 4+ DTRs in both legs. He had a negative Hoffman’s sign and 1–2 beats of clonus in both legs. He had a positive Romberg’s sign and broad-based unsteady gait.

Imaging

Initially, he had a MRI of the cervical spine without contrast [Figure 1a and b] that showed a 40 mm syrinx extending from C7 to T1. He had further workup with MRIs of the entire neuroaxis with and without contrast. A MRI of the thoracic spine with and without contrast [Figure 1d and e] showed sharp angular cord deformity at the T3 level consistent with a dorsal thoracic arachnoid web. This produced myelomalacia from T3 to T4 level with associated large intramedullary cord syrinx seen superiorly extending from the T3 through the T1 level, maximally dilated at the T2-3 level. There were rounded areas of fat signal within multiple thoracic vertebral bodies including T4, T6, T7, T10, and T11 consistent with intraosseous hemangiomas.

Surgery

The patient underwent T2-T4 laminectomies with resection of arachnoid web and use of intraoperative neuromonitoring [Figure 2]. Wide laminectomies were completed bilaterally at T2, T3, and T4, with extra caution used to keep the facet intact. There was a significant amount of fibrosis and epidural fat outside of the dura at the level of T3 that had to

be carefully reflected away to visualize the dura. Dura was opened in the midline and tacked up using 4–0 Nurolon. Microscissors and a micronerve hook were used to carefully dissect the arachnoid band off the spinal cord. This was explored laterally on both sides to ensure that there was no further tethering. At the end of the resection, the spinal cord appeared very nicely decompressed and the dura was closed with 4–0 Nurolon in a watertight fashion. A Valsalva maneuver was performed and there was no evidence of a cerebrospinal fluid (CSF) leak. Tisseel was applied and a 7 flat JP (Jackson Pratt) drain was left in the epidural space and the incision was closed in the standard fashion. Permanent pathology showed bland leptomeningeal tissue with fibrosis and calcification and no evidence of malignancy, consistent with arachnoid web.

Postoperative course

The patient was seen in clinic 2 weeks after surgery with complete resolution of his symptoms. He had returned to walking 3 miles daily by postoperative day 2. His incision was well healed; staples were removed and he was full strength on examination. He was cleared to return to work 2 weeks after that. He had a new MRI thoracic spine with and without contrast 1 month postoperative that showed complete resolution of syrinx and no further spinal cord compression [Figure 1c and f]. He was seen in clinic 1 year later for follow-up and was doing extremely well with no sequela and a repeat MRI showed no syrinx.

Case 2

History and examination

Patient is a 58-year-old gentleman who presented to the VA neurosurgery clinic with complaints of neck pain and left arm pain. His symptoms started 4 months before his clinic visit. He had left-sided neck pain and severe pain down his entire left arm into all of his fingers. He described this pain as constant with slight improvement with hot showers and taking gabapentin 1200 mg 3 times daily. He also had pain and numbness along his left thorax and left ear and face numbness. He had noted some left arm weakness. He denied any recent trauma (prior MVC 15 years ago), falls, changes in balance, problems with fine motor skills, or any bowel or bladder incontinence.

Prior medical history includes gastroesophageal reflux disease with Barrett’s esophagus, generalized anxiety disorder, recurrent major depressive episodes, hypertension, restless legs syndrome, cocaine abuse, hepatitis C, diabetes mellitus type 2, and diabetic peripheral neuropathy. He had no prior spine surgeries.

Laboratories were unremarkable. On examination, he was full strength in his right arm and both legs and was three out

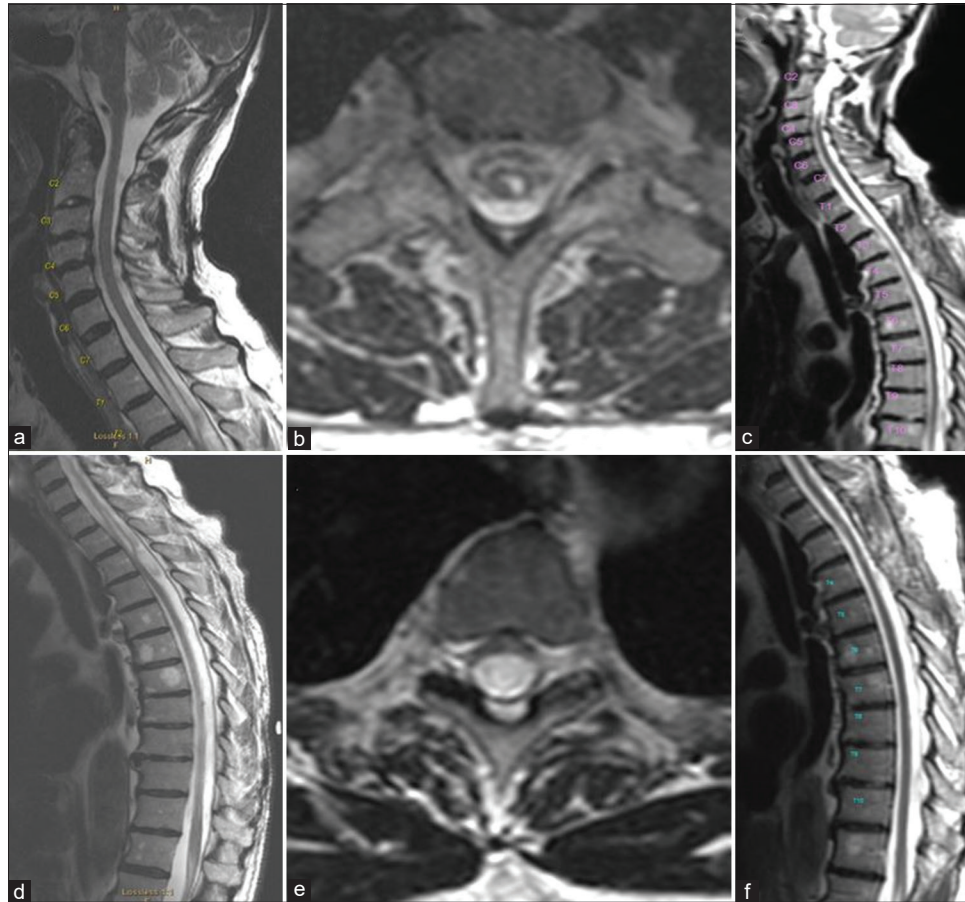


Figure 1: Preoperative (a, b, d, e) and postoperative (c, f) MR images obtained in case 1. Sagittal images (a, d) show syrinx extending up to C7-T1 with “scalpel” configuration at T3. Axial images at the level of C7-T1 (b) show syrinx and at the level of T3 (e) show severe spinal cord compression. Postoperative sagittal images (c and f) show complete resolution of syrinx and spinal cord compression.

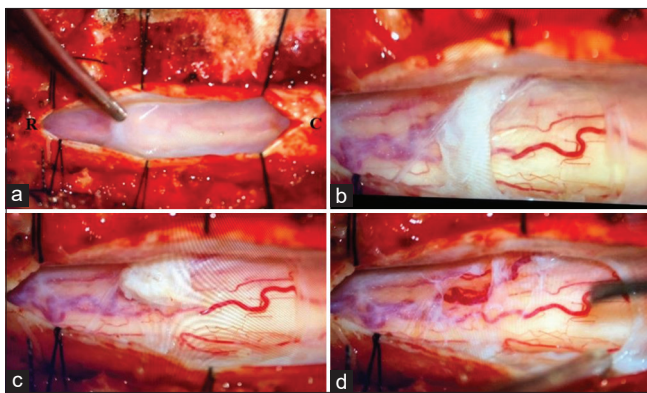


Figure 2: Intraoperative photographs obtained in case 1. (a) The thickened white arachnoid web is apparent at approximately T3 with venous congestion in the spinal cord noted rostrally. (b) Microdissection of the band from the caudal side. (c) Partial resection of thickened arachnoid web with remaining attachment on the right side (top). (d) Complete resection of arachnoid web with resolution of spinal cord venous congestion. C: Caudal, R: Rostral.

of five strength in his left deltoid and four out of five strength in the rest of his left arm. Sensation was intact to light touch

throughout, except decreased in the left arm, left hemithorax, and left jaw. He had diminished DTRs in his left arm. He had a negative Hoffman’s sign, no clonus and a negative Romberg’s sign. He had significant left arm muscle atrophy.

Imaging

He underwent a MRI of his neuroaxis including MRI head without contrast that showed no acute pathology, incidental pineal gland cyst, and mild inflammatory changes along left mastoid. His MRI of the cervical spine with and without contrast [Figure 3a and b] showed cord signal change and syrinx on the left spinal cord from C2 to C7 with syringomyelia from C7 extending into thoracic spine as well as multilevel degenerative disc disease and severe C5-C6 stenosis with cord compression. A MRI of the thoracic spine with and without contrast [Figure 3c-e] showed syrinx extending to T7-T8 measuring 9.4×7.1 mm with no enhancement, severe compression of spinal cord at T7-T8 with anterior displacement of spinal cord. A MRI of the lumbar spine with and without contrast showed

degenerative disc disease, severe L4-L5 stenosis with thecal sac compression, moderate L1-L4 stenosis, and facet hypertrophy. He underwent a diagnostic spinal angiogram which did not show any vascular abnormality or fistula and a cerebral angiogram which did not show any cervical feeders.

Surgery

The patient underwent T7-T9 laminectomies with resection of arachnoid web, midline myelotomy with drainage of intramedullary syrinx, and use of intraoperative neuromonitoring [Figure 4]. Wide laminectomies were completed bilaterally at T7, T8, and the superior aspect of T9, with extra caution used to keep the facet intact. There was a moderate amount of fibrosis outside of the dura at the level of T7-T8 that had to be carefully reflected away to visualize the dura. Dura was opened in the midline and tacked up using

4-0 Nurolon. There was a large arachnoid web band at T8 with significant tethering and compression of the spinal cord with venous congestion noted superiorly. Microscissors and a micronerve hook were used to carefully dissect the arachnoid band off the spinal cord. The web encompassed nerve roots laterally and careful attention were taken to dissect the band away from the nerves. This was explored laterally on both sides to ensure that there was no further tethering. The spinal cord was obviously expanded due to the syrinx above the level of the web and a microblade was used to make a small midline myelotomy and the syrinx cavity was visible. A small angiocath was temporarily placed through the myelotomy to allow for further drainage of the syrinx. At the end of the resection, the spinal cord appeared very nicely decompressed and the dura was closed with 4-0 Nurolon in a watertight fashion. A Valsalva maneuver was performed and there was no evidence of a CSF leak. Tisseel was applied and a 7 flat JP drain was left in the epidural space and the incision closed in the standard fashion. Pathology revealed fibrosis and arachnoid web.

Postoperative course

The patient was seen in clinic 2 weeks after surgery with significant improvement of his symptoms, especially his numbness and tingling. He continued to have persistent left arm pain, but only in a C6 distribution after surgery. His incision was well healed, staples were removed and the patient was full strength on examination. He had a new MRI of the cervical and thoracic spine with and without contrast 1 month postoperative that showed near-complete resolution of syrinx and no further spinal cord compression at the level of the arachnoid web, but remaining severe stenosis at C5-C6 [Figure 5a-d]. He underwent C5-C6 anterior cervical discectomy and fusion [Figure 5e]. He did well postoperative

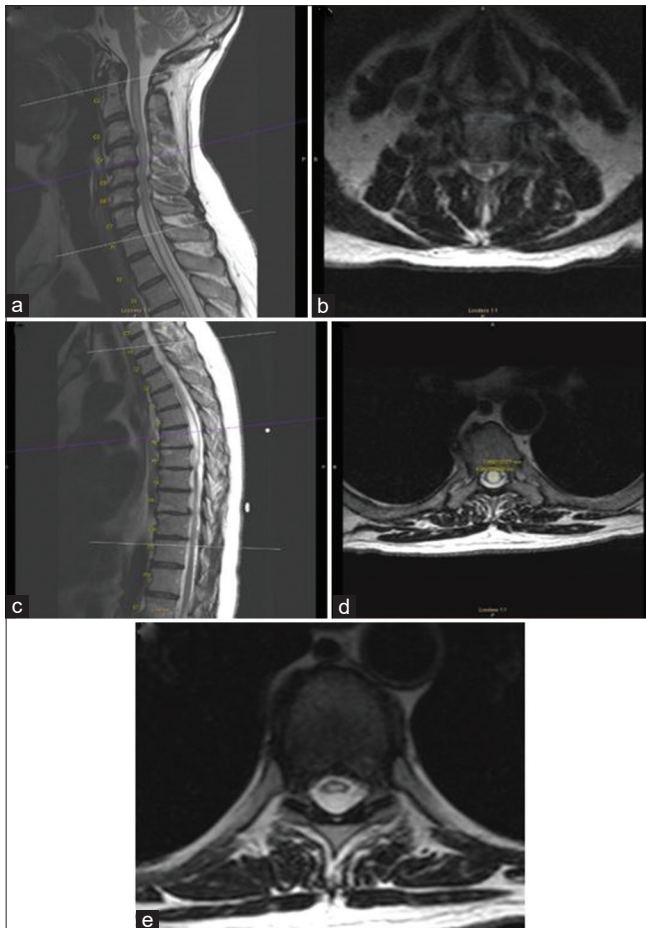


Figure 3: Preoperative MR images obtained in case 2. Sagittal and axial images (a and b) show large syrinx extending up through the cervical cord and primarily on the left side above C7. Thoracic sagittal (c) and axial images at the level of T6 (d) show large 9.4 × 7.1 mm syrinx at its maximum diameter and at the level of T7-T8 (e) show spinal cord compression due to the arachnoid web.

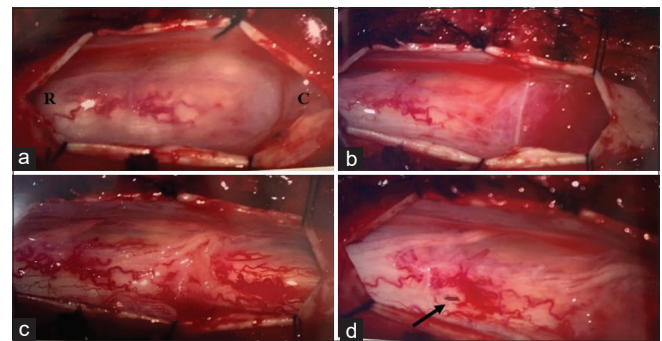


Figure 4: Intraoperative photographs obtained in case 2. (a) The cloudy thickened arachnoid web is seen caudally. (b) After slightly extending durotomy caudally, the full extent of arachnoid web is appreciated with spinal cord venous congestion inferiorly. (c) Complete resection of arachnoid bed with improvement of venous congestion. (d) Small midline myelotomy (arrow) to allow for drainage of large syrinx. C: Caudal, R: Rostral.

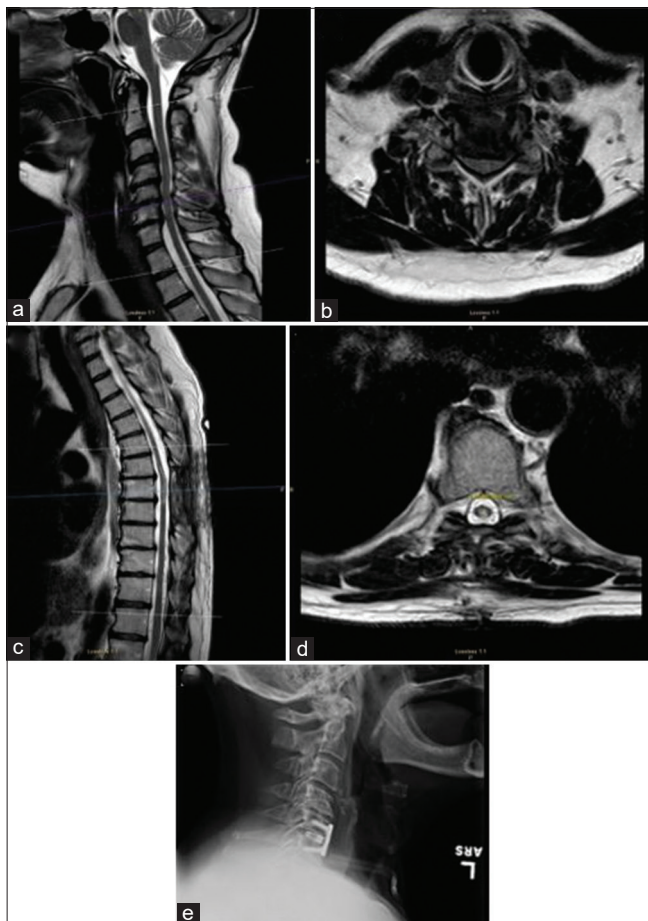


Figure 5: Postoperative MR images (a-d) and X-rays (e) obtained in case 2. Cervical sagittal (a) and axial images (b) show complete resolution of cervical syrinx with residual severe C5-C6 stenosis. Thoracic images (c and d) show near-complete resolution of syrinx and resolution of spinal cord compression. (e) Interval anterior cervical discectomy and fusion at C5-C6.

with improvement of pain, but continued to report some residual left arm numbness and pain.

DISCUSSION

Thoracic arachnoid webs are a rare entity with only 63 reports documented in the literature.^[1,3,7-10,14,15] A recent literature review that included 19 journal articles with 41 total patients showed a mean age of 52 years with diagnosis being 2.6 times more likely in males.^[10] The most common presenting symptoms are weakness (67%), sensory loss (65%), and incontinence (19%).^[10] Nearly half (47%) of patients had symptoms for 1 year or greater before surgical intervention was performed with the mean time from symptom onset to treatment of 3.3 years.^[1,3,7-10,14,15] One unique aspect with our first case report was how quickly he presented, within 2 weeks of symptom onset. Almost all of the cases reported in literature have been described as an insidious onset over months to years.

Another unique aspect of our second case was that the patient was more symptomatic from the syrinx caused by the thoracic web rather than the compression from the web. He had significant left arm atrophy and pain due to the syrinx extending into his cervical spine up to C2, which is quite a rare finding. Only three cases documenting an associated syrinx extending into the cervical spine have been reported in literature.^[10] We also decided to perform midline myelotomy at the level of the most expansile portion of the syrinx (just above the arachnoid web) to facilitate syrinx drainage. Intraoperatively, there was brisk egress of CSF from the syrinx and the expanded spinal cord decreased in size after the small myelotomy. Postoperatively, he had immediate improvement of his symptoms that were caused by his syrinx.

In reviewing the literature, all arachnoid webs were found exclusively in the thoracic spine with a syrinx associated with over two-thirds of cases.^[10] There is a clear propensity for the upper thoracic spine, ranging from T1 to T8. Surgical resection is extremely beneficial with over 90% of patients having improvement of symptoms postoperatively, making diagnosis critical in these patients.^[1,3,7-10,14,15]

Imaging diagnosis

Imaging is extremely important in diagnosing arachnoid webs, especially when differentiating arachnoid cysts and spinal cord herniation. The “scalpel sign” is a pathognomonic radiologic finding describing the CSF space outline and spinal cord indentation on sagittal imaging that is seen with arachnoid webs.^[2,4,13] One group has published on using cardiac-gated phase-contrast cine-mode MRI encompassing the area of interest and quantitatively analyzing the CSF flow.^[5] This method localizes the level of the lesion and demonstrates the one-way valve like pattern of CSF blockage. Despite this specialized type of MRI, it still may not be feasible to visualize an arachnoid web on preoperative imaging due to the thin nature of the membranes. Therefore, a very thorough clinical history and examination is especially important in these cases.

Pathophysiology

Arachnoid web is an extramedullary transverse band of arachnoid tissue that extends to the dorsal surface of the spinal cord resulting in mass effect and dorsal indentation of the spinal cord.^[6] It usually is associated with syringomyelia above and/or below the level due to abnormal blockage of CSF flow, as has been well described by the intramedullary pulse pressure theory.^[6,10,13] Prior case reports have hypothesized that arachnoid webs are a subtype of arachnoid cyst that is either precursors that

have not fully walled off or the remnants of a previous arachnoid cyst that ruptured.^[11] However, no definite conclusion has been made. The histopathology of arachnoid webs has shown connective tissue with a small number of CD3+ T-cells and asymptomatic small amount of ossification. This suggests the possibility of an inflammatory process.^[5,9]

Treatment and outcomes

The most common surgical treatment is with laminectomy with intradural excision of the arachnoid web including variations of this with hemilaminectomy and minimally invasive techniques.^[10] Less commonly shunt and stent placement have been used for CSF flow diversion. There was a recent case report published of percutaneous fenestration of a thoracic arachnoid web using a standard lumbar drainage catheter with placement in the lumbar thecal sac and advancement of the catheter to the level of the arachnoid web with resultant clinical improvement.^[12] Outcomes are generally very good with treatment. In a literature review including 19 articles, patients who were followed for an average of 9.2 months had 91% improvement in neurological symptoms with only two patients having worsened symptoms. These two cases had new symptoms after syringopleural shunt placement with the development of paraplegia in one case and numbness in the right leg for the other.^[10,15] To the best of our knowledge, there are no case reports of recurrence of arachnoid webs suggesting long-term disease control after surgery.

CONCLUSION

Thoracic arachnoid webs remain a rare entity that can have severe neurological impairments with rapid improvement of symptoms postoperatively. For patients that are symptomatic from an associated syrinx, a midline myelotomy to facilitate drainage can be considered to be done concomitantly with the arachnoid web resection.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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