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Journal of Craniovertebral Junction and Spine

Case Report

Syringomyelia secondary to "occult" dorsal arachnoid webs: Report of two cases with review of literature

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Journal of Craniovertebral Junction and Spine 2016, 7:22

Abstract

In a certain group of patients with syringomyelia, even with the advent of sophisticated magnetic resonance imaging (MRI), no associated abnormality or cerebrospinal fluid (CSF) block is easily identified. This type of syringomyelia is often termed idiopathic. Current literature has less than 10 reports of arachnoid webs to be the causative factor. We present our experience in the management of two cases of syringomyelia secondary to arachnoid webs. Both our patients presented with progressive neurological deterioration with MRI scans demonstrating cervical/thoracic syrinx without Chiari malformation or low-lying cord. There was no history of previous meningitis or trauma. Both patients underwent myelography that demonstrated dorsal flow block implying CSF obstruction. Cord displacement/change in caliber was also noted and this was not evident on MRI scans. Both patients underwent thoracic laminectomy. After opening the dura, thickened/abnormal arachnoid tissue was found that was resected thus widely communicating the dorsal subarachnoid space. Postoperatively at 6 months, both patients had significant symptomatic improvement with follow-up MRI scans demonstrating significant resolution of the syrinx. In patients with presumed idiopathic syringomyelia, imaging studies should be closely inspected for the presence of a transverse arachnoid web. We believe that all patients with idiopathic symptomatic syringomyelia should have MRI CSF flow studies and/or computed tomography (CT) myelography to identify such arachnoid abnormalities that are often underdiagnosed. Subsequent surgery should be directed at the establishment of normal CSF flow by laminectomy and excision of the offending arachnoid tissue.

Key words: Arachnoid webs, idiopathic, syringomyelia

INTRODUCTION

Syringomyelia is the development of a fluid-filled cavity within the spinal cord. Despite different etiologies that cause syringomyelia, it is largely agreed that the alteration of cerebrospinal fluid (CSF) flow dynamics plays an important role in the development of a syrinx. Known causes of syringomyelia include Chiari 1 malformation and basal arachnoiditis [postinfectious, inflammatory, postirradiation, blood in subarachnoid space

Access this article online	
Quick Response Code:	Website: www.jcvjs.com
	DOI: 10.4103/0974-8237.181862

How to cite this article: Sayal PP, Zafar A, Carroll TA. Syringomyelia secondary to "occult" dorsal arachnoid webs: Report of two cases with review of literature. J Craniovert Jun Spine 2016;7:101-4.

(SAS) amongst others]. Primary spinal syringomyelia refers to syrinx formation due to a spinal pathology, e.g., spinal cord tumor, spinal infection, trauma, and surgery.

The term "idiopathic syringomyelia" has been used to describe syrinx formation in cases where causative mechanism remains undetermined after clinical and routine radiological work-up. The term does not imply the absence of a cause but just the absence of a cause that can be detected using standard diagnostic methods.

There are very few reports in the literature describing arachnoid bands/webs as causes of "idiopathic syringomyelia." We present our experience in the management of cases of syringomyelia due to dorsal spinal arachnoid bands and review the current available evidence of the diagnostic work-up and management of this etiology.

CASE REPORTS

Case 1

A 43-year-old male was referred with a 2-year history of progressive dissociative sensory loss in his lower limbs with myoclonic twitches. This was associated with neck and low back pain in addition to left-sided radiating leg pain. There was no history of previous trauma, central nervous system (CNS) infection, or spinal surgery. On examination, there was non-dermatomal numbness in left lower limb but normal power in all four limbs. He had been investigated previously in another neurosurgical center and the magnetic resonance imaging (MRI) scan revealed a cervicothoracic syrinx but in the absence of any apparent cause; the patient was managed conservatively for 2 years till he was eventually referred to our center for a second opinion.

MRI scan [Figure 1] showed twin syrinx cavities in the cervical and upper thoracic regions with an unusual distribution and no apparent etiology. The patient underwent a computed tomography (CT) myelogram [Figure 2] that showed CSF block at T1/T2 suggestive of arachnoid pathology. During the CT myelogram, a small air bubble was inadvertently introduced that ascended superiorly and was trapped at the level of the CSF block. The MRI scan had not shown any apparent abnormality at this level. He was now experiencing increasing dysesthetic pain down both arms along with left flank numbness and gait disturbance. In view of progressive deterioration, he underwent a T1/T2 thoracic laminectomy for intradural exploration that revealed a thickened dorsal band of arachnoid. This tissue was excised, and intraoperatively improved craniocaudal CSF flow was noted. The histology of the tissue revealed arachnoidal tissue.

Postoperatively, the patient recovered well from the operation and there was gradual improvement in his symptoms with normal gait and return of normal function of hands. Six months after the surgery, the patient reported complete symptom resolution and follow-up MRI scan [Figure 1] showed significant reduction in the size of the syrinx.

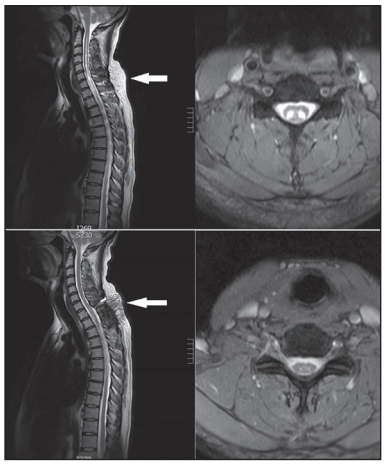

Figure 1: Preoperative (top row) and postoperative (bottom row) MRI for Case 1 showing cervical significant syrinx prior to surgery and collapsed down following surgery. The postoperative scan was done 6 months after the surgery

Case 2

A 45-year-old male was referred following an episode of legs giving way that was followed by paraparesis for a few hours that recovered fully. On examination, there was no neurological deficit.

MRI scan [Figure 3] revealed a lower thoracic syrinx with some signal change in spinal cord. There was no associated Chiari malformation or tethered cord. Follow-up imaging after 18 months showed no change. However, 4 months later the patient reported increasing discomfort and numbness in both the arms with subjective lower limb weakness and gait disturbance. A CT myelogram [Figure 4] was performed that revealed displacement of the spinal cord at T3 to T5 and compression to the left likely secondary to either arachnoid band or cyst. This had not been evident on the MRI.

By now, the patient had developed right lower limb spastic weakness along with left radiating leg pain. On examination he had brisk reflexes, positive Hoffmann's sign bilaterally, positive ankle clonus, reduced sensation in the right leg, and upgoing plantars.

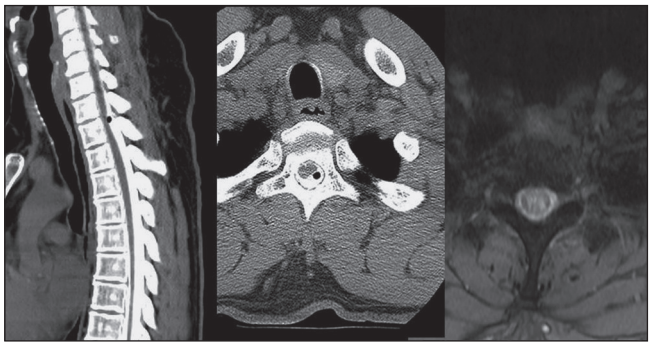

Figure 2: Preoperative CT myelogram and axial MRI for Case 1. The abrupt change in cord caliber/displacement demonstrated by the myelogram was not evident on MRI studies. Note inadvertent bubble of air that ascended superiorly to the level of arachnoid web, which was useful in confirming the presence and level of the web

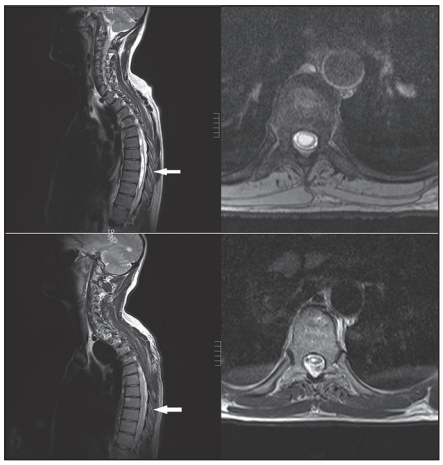

Figure 3: Pre- (top row) and Postoperative (bottom row) MRI for Case 2 showing significant syrinx prior to surgery and collapsed down following surgery.The postoperative scan was done 12 months after the surgery

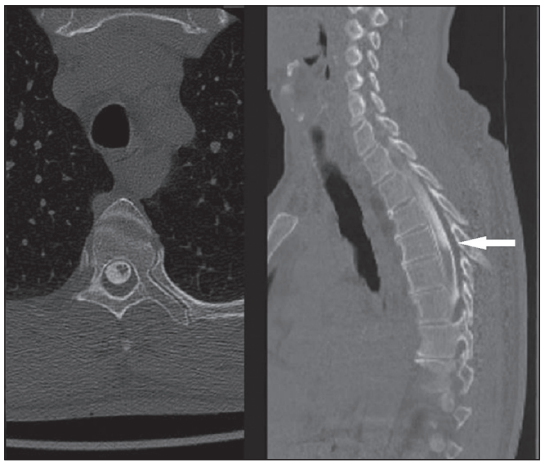

Figure 4: Preoperative CT myelogram for Case 2 demonstrating the cord displacement and change in caliber

We performed a T3-5 laminectomy. Intradurally, we found a dorsal arachnoid band that was excised and histology confirmed arachnoid tissue. Postoperatively, the patient recovered well from the operation with improvement in right leg strength and the left leg pain. At 6-month follow-up, he reported complete symptomatic resolution, and neurological examination was normal. His follow-up MRI [Figure 3] at 6 months postoperatively revealed significant reduction in the syrinx size.

DISCUSSION

We report a very uncommon and hence under-recognized cause of symptomatic syringomyelia. Whilst arachnoid bands/webs have previously been reported as the etiologic factor for idiopathic syringomyelia, it remains a rare diagnosis with very few case series reported.[1-5] Mallucci *et al*.[1] first reported the importance of recognizing occult arachnoid webs, pouches, and cysts in idiopathic syringomyelia. In a large series of 28 patients with thoracic arachnoid pathology, 6 cases of ventral or dorsal arachnoid bands were reported.[5] However, there is uncertainty not only regarding the pathophysiology of syrinx formation with arachnoid webs but also regarding the ideal radiological investigations.

Arachnoid adhesions due to trauma/infection/previous surgery are well-recognized causes of syringomyelia. However, none of our patients had these contributing factors in their history. Idiopathic arachnoid bands have been hypothesized to develop from the septum posticum, the membrane dividing the midline dorsal arachnoid space of the cervicothoracic spinal canal.[2] Alternatively, Paramour[4] hypothesized that these webs are simple variants of arachnoid cysts. It is however equally possible that they represent localized arachnoiditis due to factors that are not always elicited in the history.

Frequently cited theories in syrinx formation are those of Gardner, Williams, and Oldfield.[6-8] These theories largely focus on impaired CSF flow at the foramen magnum. However, these theories do not address primary spinal syringomyelia. Greitz[9] proposed that increased intramedullary pulse pressure in the spinal cord relative to the nearby SAS is the distending force in the production of syringomyelia and that syrinx consists of extracellular fluid accumulated in the distended spinal cord.

Heiss *et al*.[10] directly addressed the pathophysiology of primary spinal syringomyelia in their prospective study of 36 patients. Their findings were consistent with their theory that a spinal subarachnoid block increases subarachnoid pulse pressure above the block, producing a pressure differential across the obstructed segment of the SAS, which results in syrinx formation and progression. They reported that their findings were similar to that of their previous studies, which examined the pathophysiology of syringomyelia associated with obstruction of SAS at the foramen magnum, inferring a common mechanism for both. Three-dimensional computational models of the spinal SAS have been used to study CSF flow either in unobstructed SAS or with SAS obstructed by a porous region simulating dorsal and circumferential arachnoiditis.[11] The findings suggest that syrinx formation may be related to a change of temporal CSF pulse pressure dynamics. Consequently, it would appear that restoring CSF flow across the arachnoid webs/bands should result in syrinx resolution. Alternatively, syrinx formation may occur due to resonance in a mechanical system with a pathology-induced nodal point.

Arachnoid webs are difficult to diagnose using conventional MRI imaging. However, the recognition of a secondary imaging finding "the scalpel sign" can suggest the presence of an arachnoid web. This is a focal indentation of the thoracic spinal cord, resembling a scalpel on sagittal MRI images with the scalpel blade pointing posteriorly.[12] However, none of our patients had any clear evidence suggesting an arachnoid web on conventional MRI.

In all the early reported series of arachnoid webs, CT myelography was the investigation of choice. Indeed, it has been reported that cardiac gated MRI techniques were less successful in studying CSF flow in the spine than at the craniovertebral junction.[13] However, in a large series of 125 patients with idiopathic syringomyelia, Mauer *et al*.[14] performed cardiac gated phase contrast MRI of

CSF flow in the median sagittal plane in the spine and reported that these were more reliable compared to myelography. Cardiac gated phase contrast cine-mode MRI in multiple axial planes has also been used to quantitatively analyze CSF flow in those planes for the diagnosis of arachnoid webs.[15] Whilst in these papers, arachnoid webs were indirectly diagnosed using flow studies; direct visualization of arachnoid membranes has been reported with high resolution using retrospectively cardiac gated cine steady-state free precession MRI.[16] However, all these MRI techniques are susceptible to movement artifacts and also require a 3Tesla MRI that may not always be available. Arachnoid webs may be missed even with MRI CSF flow sequences, hence CT myelography remains useful. It is worth noting that the inadvertent air bubble introduced during the CT myelogram of our first patient provided an excellent clue about the location of the CSF block.

The location of the syrinx is not a reliable clue regarding the level of arachnoid web/CSF block.[14] The arachnoid web was just caudal to the syrinx in our first patient and at the cranial end of the syrinx in the second patient.

Both our cases were reliably diagnosed using CT myelography. We recognize that myelography is an invasive procedure with potential morbidity and with recent significant advancements in MR technology, it may be prudent to investigate symptomatic idiopathic syringomyelia with MRI CSF flow studies in the first instance and failing that, to proceed to myelography.

We believe that restoring CSF flow by excision of the arachnoid band and thereby improving SAS compliance offers the best possible outcome in these symptomatic cases. In the earliest reported series, all patients treated by web excision improved whereas both patients treated with syringo-pleural shunts worsened.[1] Mixed result were obtained with shunting in other series but excision of the webs was almost always associated with successful outcome.[2,3,5] Shunting syrinxes is associated with high rates of recurrence, poor long-term outcomes, and sometimes worsening of neurological status.[1,3,13] In fact, most patients with any short segment arachnoid pathology causing primary spinal syringomyelia, generally have good outcomes after surgical decompression/detethering procedures. It is the patient group with long segment pathology that often requires multiple surgeries, frequently for CSF diversion. This was corroborated in a recent systematic review, which suggested that arachnolysis rather than shunting was the only initial surgical treatment that is likely to have a statistically significant effect on lowering recurrence rates.[17] This once again highlights the importance of exhaustively looking for a rectifiable surgical targets, such as arachnoid webs and cysts in the presumed idiopathic syrinxes, which would otherwise have be treated with shunts.

CONCLUSION

Arachnoid webs/bands are a rare and hence often underdiagnosed cause of idiopathic syringomyelia, leading to delayed diagnosis and treatment. In symptomatic/deteriorating patients with idiopathic syringomyelia, imaging studies should be closely inspected for the presence of a transverse arachnoid web. Conventional MRI should always be augmented with MRI CSF flow studies in the sagittal plane and if needed in multiple axial planes to aid in the diagnosis. CT myelography should be performed if these studies do not yield answers or also if the surgeon feels further information is needed prior to definitive treatment, especially as myelography provides more robust localization of the level of block. Focussed surgical decompression and excision of the offending arachnoid tissue usually leads to good clinical and radiological outcomes.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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