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Case Report

Extensive spinal intradural arachnoid cyst exhibiting a “double cord sign” on magnetic resonance imaging

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ABSTRACT

Study design: Retrospective case study.**Objective:** To report on a case with an extensive intradural arachnoid cyst that caused left hemiparesis.**Summary of background data:** Intradural arachnoid cysts are not common causes of spinal cord compression and myelopathy. Although arachnoid cysts of the spine have been occasionally reported, cases with extensive intradural arachnoid cysts associated with hemiparesis are rarely reported.**Method:** The patient was a 43-year-old woman who had cervical myelopathy symptoms, including radiating pain and left limb hemiparesis with gait disturbance. Magnetic resonance imaging revealed an extensive intradural extramedullary multi-septated cyst from the C6–T12, exhibiting a double cord sign on the T2-weighted axial image of the spine. The mass blocked normal cerebrospinal fluid flow, which led to cord compression. Two stages of operations were planned because of the patient's health status. The patient underwent laminectomy and cyst wall resection on the level of the T5–T7, which had the most compressed lesion. From C6–T2, a right side unilateral laminectomy was performed to resolve the right limb's neurology after the first operation.**Result:** Hypoesthesia and the radiating pain were improved immediately postoperatively; however, motor power at the C8–T1 level of the right unaffected side was impaired and included finger abduction and finger flexion, which decreased to 4/5. During the second operation, the arachnoid cyst was exposed by performing a right unilateral laminectomy. The patient's neurologic symptoms were improved without neurologic sequelae.**Conclusion:** In the case of an extensive cyst that exhibits a double cord sign, an intradural arachnoid cyst should be suspected as a differential diagnosis. Without radical excision of all lesions, neurologic symptoms can be recovered by performing selective resection and CSF normalization can be achieved even at extensive levels.

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1. Introduction

Although arachnoid cysts of the spine are occasionally reported, cases with extensive intradural arachnoid cysts associated with hemiparesis are rarely reported. Intradural arachnoid cysts of the spine are uncommon lesions¹ that cause spinal cord compression and are described by other symptoms.² In most cases, acquired arachnoid cysts in adults caused by infection, cord trauma, or other mechanisms result in inflammation and subarachnoid adhesions.³⁻⁶ Arachnoid cysts are usually located in the mid-thoracic spine to the lower thoracic spine, are more rare in cervical lesions, are mainly in a posterior position instead of an anterior position,⁷ tend to be symptomatic during the second decade of life,² and are more common in males than females.⁸ Many theories have been suggested to explain the cyst formation and mechanism of symptoms. Perret et al proposed that they are caused by diverticulum in the septum.^{6,9} Fortuna et al^{10,11} suggested that strangulation of the arachnoid granulations can produce cerebral spinal fluid (CSF), which becomes blocked by the arachnoid diverticulum. These fluid sequestrations lead to further disruption of the normal CSF flow, which can expand and cause syringomyelia. The clinical manifestations of arachnoid cysts are similar to those of cervical myelopathy, such as paresis, radicular pain, paresthesia, and gait disturbance. Headaches can also be the main symptom in patients with a cervical arachnoid cysts, while some experience urinary incontinence.¹² According to other authors, severe flattening of the spinal cord due to a cyst and a long-term duration of paresis are predictors of a poor outcome, especially in the elderly.¹³ The treatment of choice is complete cyst removal^{2,6,12}; however, if this is not possible, partial resection in the affected levels can be performed.¹² We report on a case with an extensive intradural arachnoid cyst, which had a double cord sign on magnetic resonance imaging (MRI) and was associated with left hemiparesis.

2. Case report

2.1. Presentation and examination

A 43-year-old woman presented with a history of 6 months of hypoesthesia and gait disturbance and radiating pain in the left limbs, without any previous history of trauma or infection. She underwent non-surgical physiotherapy for 6 months without any improvement. Patient had gait disturbance in addition to difficulty in controlling her sphincter reflexes that modulate defecation and micturition. The physical examination demonstrated motor power with a 4/5 strength grade for all muscle groups in the left extremities, and the knees had hyper-reflexive responses. The sensory examination showed reduced proprioception and vibratory sensation in her left limbs. Computed tomography (CT) of the whole spine showed thinned pedicles, an increased diameter of the spinal canal, and a posterior scalloping distortion of the vertebral bodies, especially from C6–T12. Preoperative MRI demonstrated an extensive posterior intradural extramedullary arachnoid cyst from the C6–T12 vertebral bodies (Fig. 1). The spinal cord was

compressed and flattened anteriorly. From T6–T8, the shape of the cyst, which faced the cord, appeared as if it had a stack of circles (i.e., a double cord sign) (Fig. 2).

2.2. Surgical procedure

Since the cystic structure was extended considerably from C6–T12 and considering the condition of the anemic patient, a staged operation was scheduled to be performed sequentially within 1 week. The patient was planned to undergo the first operation on the T5–T9 lesion, which was the most compressed location. General anesthesia was administered, and the patient was in the prone position. Continuous electrophysiologic monitoring for somatosensory and motor evoked potentials was utilized in our case to monitor any neurologic changes that might occur during the operation. Total laminectomy was performed from T5–T8 to expose the arachnoid cyst. As soon as the laminectomy was complete, a thin-walled expended dura, including the cystic structure occupying the canal, was found. Prior to cyst removal, durotomy was performed using a knife and Pott scissor. The cystic fluid was colorless, the CSF had accumulated in each septum so that the normal flow of CSF was not observed, and the cord was also compressed with considerable pressure caused by the septated cystic structure (Fig. 3). The cystic structure was

Fig. 1 – Preoperative sagittal T2-weighted magnetic resonance imaging (MRI) of the spinal column. It demonstrates cord compression and the intramedullary arachnoid cyst extending from C6 to T9.

Fig. 2 – Preoperative axial T2-weighted magnetic resonance imaging at the T8 level. The arrow indicates spinal cord compression by the septated arachnoid cyst that is filled with cerebral spinal fluid. It is possible to differentiate between the cyst and the intradural space, and it can be depicted as a double cord sign.

separated into several septa that blocked normal communication. After the dura was opened, the cyst wall was resected with fenestration to the subarachnoid space. The meticulous dissection of any adhesions was performed to untether the spinal cord. Finally, the cystic structure was removed until the normal CSF flow was visible. After verifying the CSF flow in the spinal cord, the dural defect was repaired using 6-0 prolene sutures and by applying collagen fleece and fibrin glue. After dural repair, no further CSF leakage was identified via the Valsalva maneuver. The cyst wall revealed a pathological diagnosis of an arachnoid cyst. The postoperative MRI evaluation performed at 1 week following the surgery revealed the decreased size of the

Fig. 3 – An intraoperative photograph. The dural sac is opened and retracted by sutures. Compression of the spinal cord by the arachnoid cyst and disturbance of the normal cerebrospinal fluid flow is observed.

arachnoid cyst at the T4–T9 level along with decompression of the flattened spinal cord (Fig. 4). Hypoesthesia and the radiating pain were considerably improved immediately post-operatively; however, impairment of motor power at the C8–T1 level occurred and the motor power of the right unaffected side preoperatively, finger abduction, and finger flexion decreased to 4/5 compared with the previous symptoms. During the second operation, the arachnoid cyst was exposed by performing a right unilateral laminectomy, because the patient complained about her right upper limb pain and weakness related to the C8–T1 nerve where compression was observed on the postoperative MRI. Dissection of any arachnoid adhesions was performed, and the cystic structure was removed until the normal CSF started to flow as observed in the preceding operation. The histopathologic study of the cyst wall revealed fibrous connective tissue with an inner single cell lining (Fig. 5). The patient's neurologic symptoms were improved without any neurologic sequelae.

Fig. 4 – Postoperative sagittal T2-weighted magnetic resonance imaging (MRI) of the thoracic spine. The arachnoid cyst is excised, and spinal cord diameter has recovered in comparison to the previous MRI.

Fig. 5 – Microscopy of the specimen demonstrates the cyst wall, which consists of fibrous connective tissue with an inner single cell lining (hematoxylin-eosinophil stain; high power field, $\times 400$).

3. Discussion

Spinal arachnoid cysts are relatively rare, accounting for 1% of all spinal tumors.⁸ They have been related with abnormalities of the spine due to trauma, hemorrhage, infection, neural tube defects, and spinal deformity deformities.^{4,14,15} Our case is unique, because the patient's spinal arachnoid cyst was not caused by any anomalies, and it exhibited an extensive lesion with a double cord sign on the axial MRI of the spine. While Kumar et al¹⁶ explained the mechanism of cyst enlargement, there are several theories postulated by other authors, which include the following: 1) secretion of the cyst cell; 2) existence of a one-way valve; and 3) the pathological disposition of an arachnoid trabecule caused by diverticulum. In our case, it is difficult to know which mechanisms are associated with the provocation of symptoms among these factors. During the surgery, we found a CSF pouch consisting of trabeculation and multiple septation, which blocked the normal CSF flow. It is possible to assume that an etiopathogenic mechanism cannot be explained by only one theory. The cyst expanded from C6–T12 through many vertebrae, even though rare data were found associated with the incidence of cysts and their location. A literature review on the specified location of arachnoid cysts indicated that the mid-thoracic spine has the highest incidence among other vertebra for lesions, whereas the incidence for lesions on the cervical spine is relatively rare; however, a posterior cyst is more common than an anterior cyst.^{6,10,11,13,14,17} Wang et al⁶ postulated that the signs and symptoms of spinal arachnoid cysts are attributed to the chronic cyst expansion and the compression of the neural elements, while the preoperative symptoms and signs may be different according to the location of the cystic structure and compression of the spinal tracts. Anterior cysts tend to cause weakness and myelopathy, whereas dorsal cysts are present more commonly with neuropathic pain and numbness. In our case, the patient complained of radiating pain and dominant numbness, including myelopathy symptoms, such as gait

disturbance and difficulty with defecation and micturition. MRI is critical for the exact diagnostic evaluation of arachnoid cysts.^{6,12,18} MRI can show how many levels of vertebrae are affected and where the cyst is compressing the neural element. This tool enables surgeons to resect the cyst accurately and to decompress the involved levels and prevent unnecessary laminectomy. Upon reviewing other cases, it was apparent that an arachnoid cyst involving several vertebrae was occasionally reported. However, a long extensive intradural arachnoid cyst from the cervical to the upper lumbar spine with distinctive MRI findings of a circle stacked on a circle (i.e., the double cord sign) was rarely reported, to our knowledge. The cysts were non-communicating through the subarachnoid space. The cyst led to a blockage in the normal CSF flow, resulting in epidural cystic enlargement contributable to spinal compression. As the cyst expanded into the limited canal space, the cyst was confronted with the cord and was visualized on an axial MRI scan. An intradural arachnoid cyst should be differentially diagnosed if a double cord sign is observed on an axial MRI scan. The treatment of a subarachnoid cyst has not been standardized. Many authors propose surgery, including complete resection of the cyst wall and closure of the communication site between the cyst and the subarachnoid space after laminectomy of the affected vertebra.^{2,6,7,12,19} If complete resection is impossible, other methods are also available as adjuvant procedures, such as fenestration of the cyst wall, percutaneous drainage, or cyst shunting to the atrium, pleura, or peritoneum.^{10,16,20–22} If the cyst adheres to the spinal canal, marsupialization is recommended to relieve symptoms successfully.²³ The best reliable treatment is complete surgical excision of the cystic structure in order to recover the neurologic symptoms. In our case, because the lesions were long extending arachnoid cysts, it is difficult to determine how many levels should be performed during the operation. Thus, the patient underwent staged operation sequentially to prevent any damage, such as a kyphotic deformity. Although the cyst could not be completely resected due to technical difficulty, we made a great effort to remove the cystic structure and to open the communication of CSF flow throughout the surgery. The flow of CSF should be clearly identified several times via the Valsalva maneuver before repairing the dura. In the case of a long lesion extending through many vertebrae, care should be taken to not choose laminectomy, as it would negatively influence the spinal column stability.¹² During the first operation, there was a relatively lower possibility to cause severe kyphotic deformity, as cyst removal was performed by laminectomy of the T5–T8 level. Although the previous left side symptoms (i.e., the radiating pain and hypoesthesia) were improved, a new problem arose postoperatively, because the patient had radiating pain and finger abduction weakness on the opposite side. Postoperative MRI was performed at 5 days, and the cystic structure had deviated to the right side, especially from C7–T1 (Fig. 6). Because the literature is lacking on patients who developed radiating pain on the contralateral side and weakness after undergoing decompression of the arachnoid cyst, it was impossible to know why the patient developed contralateral radiating pain even though her left side neurologic symptoms were recovered. We assumed that the compensation mechanism resulted in right subdural space

Fig. 6 – Postoperative axial T2-weighted magnetic resonance imaging at the T1 level. The arrow shows the cord compression caused by the right side of the cyst.

expansion after the first operation in which we excised the cystic structure dominantly on the left side. After recovery of CSF flow at the most compressive lesion, it was presumed that the flow migrated into the right side of the cyst (i.e., the C7–T1 lesion). During the second operation, if the patient underwent multilevel laminectomy instead of a unilateral minimal laminotomy to remove the cysts, there was a high possibility of causing severe kyphotic deformity. Since the cervicothoracic junction can lead to kyphotic deformity after multilevel laminectomy, we performed obliteration of all the cystic cavities and decompressed the neural element copiously to recover free-flowing CSF by unilateral laminotomy. The patient's symptoms, including the radiating pain and motor weakness, were improved in comparison to the preoperative state.

4. Conclusion

In the case of an extensive cyst that exhibits a double cord sign on an axial MRI of the spine, an intradural arachnoid cyst should be suspected as a differential diagnosis. Without radical excision of all lesions, the neurologic symptoms can be recovered by selective resection and CSF normalization, even if the cyst extends to several levels.

Conflicts of interest

All authors have none to declare.

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