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Giant Craniospinal Intramedullary Arachnoid Cyst: A Rare Occurrence

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A spinal arachnoid cyst can be extradural, intradural, or intramedullary. Intramedullary arachnoid cysts are rare and more common in the thoracic spinal cord. Cervical intramedullary arachnoid cysts are extremely rare, and only few cases are reported in the literature.[1,2]

Literature search do not reveal such giant craniospinal intramedullary arachnoid cyst.

A 45-year-female presented with difficulty in walking, tightness, and sensory symptoms in all four limbs. Her neurological examination revealed hypertonia in all four limbs with exaggerated deep tendon reflexes in all four both limbs and superficial reflexes were absent. Babinski was positive on both sides. Her recent magnetic resonance imaging (MRI) of cervical spine revealed a well-defined, purely cystic lesion extending from the pontomedullary junction to the cervical spinal cord up to the C7 level that was approximately $10.1 \times 3.0 \times 2.6$ cm in size. The lesion was homogenous hypointense on T1-weighted images, hyperintense T2-weighted images, and not enhancing with intravenous gadolinium administration (►**Fig. 1A–D**). Earlier she was treated twice in the form of needle aspiration with recurrence in 4 months. She underwent excision of the arachnoid cyst under general anesthesia in the prone position. Cervical laminectomy was performed from C1 to C5. Dura opened in the midline from C1 to C7. The tense spinal cord was visualized with a translucent fluid-filled cystic lesion at the spinal cord's upper end (►**Fig. 1E**). The cystic lesion was entered via midline myelotomy of about 1 cm, and clear translucent water-like fluid drained out. A similar myelotomy was performed at the lower end of the cervical spinal cord. There was no exact cleavage plane between the cyst wall and spinal cord parenchyma. Part of the translucent membrane was biopsied and sent for histopathological examination. At both myelotomy sites, a 1 cm size piece of silastic tube was anchored in such a way so that it communicates with intracystic space and extramedullary subarachnoid space (►**Fig. 1F**). Primary water-tight dural closure ensured. Wound closed in layers. The postoperative course was uneventful. She had significant improvement in her neurological symptoms. Histopathological examination of the cyst wall revealed evidence of an intramedullary arachnid cyst (►**Fig. 2**). At 4 years following, she had no clinical symptoms. Her neurological examination revealed no abnormality. Follow-up cervical spine MRI showed near-complete resolution of the intramedullary cystic lesion with no recurrence evidence (►**Fig. 1G–I**).

The natural history of this extremely rare pathology is unclear. Asymptomatic cysts can be observed while symptomatic cysts should be treated. The most effective treatment option in the case of symptomatic intramedullary arachnoid cyst is debatable.[3] Surgical excision or fenestration of the cyst is the treatment of choice. Laminectomy or laminoplasty is performed, a cyst is exposed and fenestrated or marsupialized if total excision is not possible by midline myelotomy or, in some cases, via dorsal root entry zone myelotomy.[4] No worsening of symptoms was observed in either of the approaches.[3] Like neuroepithelial cyst, arachnoid cyst does not have a clear plane of cleavage.[5] As the cyst wall is a continuation of spinal cord parenchyma, one should not separate it. Widening of the spinal cord opening and marsupialization of the cyst result in significant neurological improvement.[1,2] Compared with the extramedullary location, complete removal of the intramedullary cyst is not possible, and shunt placement is required.[6] Cystoperitoneal shunt is considered in refractory cases.[7]

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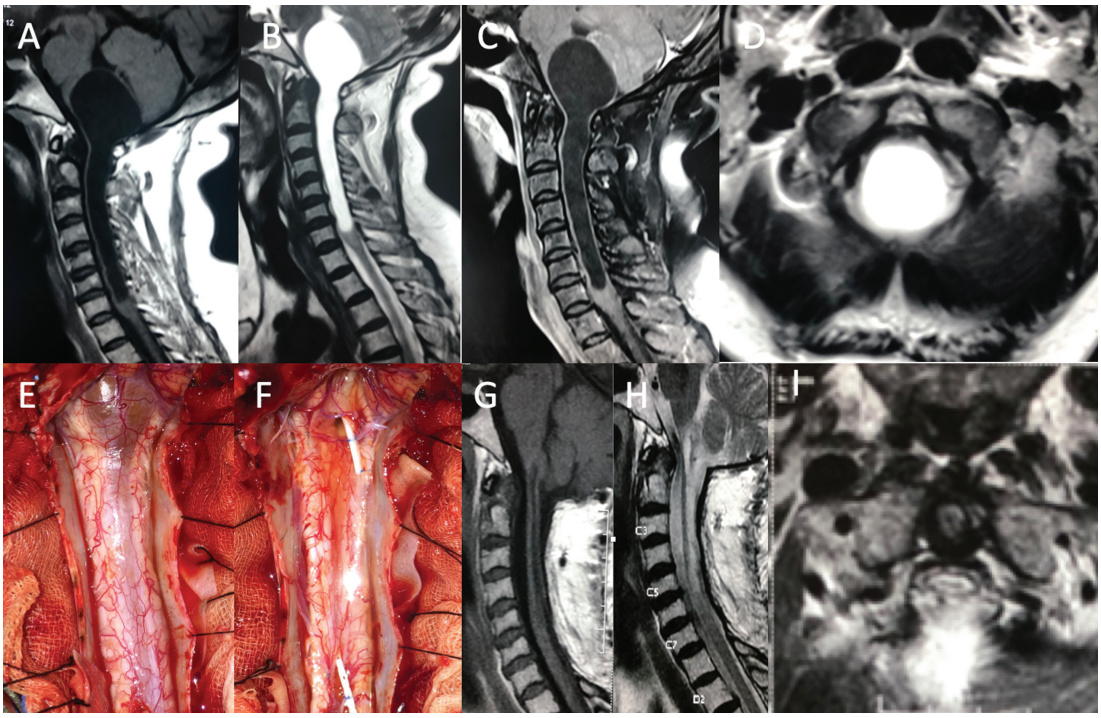

Fig. 1 Magnetic resonance imaging (MRI) of the craniovertebral junction with cervical spinal (sagittal view) showing well-defined, purely cystic lesion extending from pontomedullary junction to cervical spinal cord up to C7 level. The pontine end appeared bulbous or ice cream on the cone, while the cervicomedullary portion seemed to be pipe-like or cone of ice cream. The lesion also appeared homogenous hypointense on T1-weighted images (**A**), hyperintense on T2-weighted images (**B**), and not enhancing on intravenous gadolinium administration (**C**). MRI high cervical spine axial view showing cystic lesion on T2-weighted images (**D**). Intraoperative photomicrograph showing expanded spinal cord with a surface extending translucent lesion mainly at the cervicomedullary junction (**E**) and decompressed spinal cord with small myelotomy and cysto-subarachnoid shunt at the upper and lower end of the lesion (**F**). Four-year follow-up MRI of the craniovertebral junction with cervical spinal (sagittal view) showing near-complete resolution of lesion on T1-weighted (**G**) and T2-weighted images (**H**). Axial view of the upper cervical spine showing near-complete resolution of lesion on T2-weighted images (**I**).

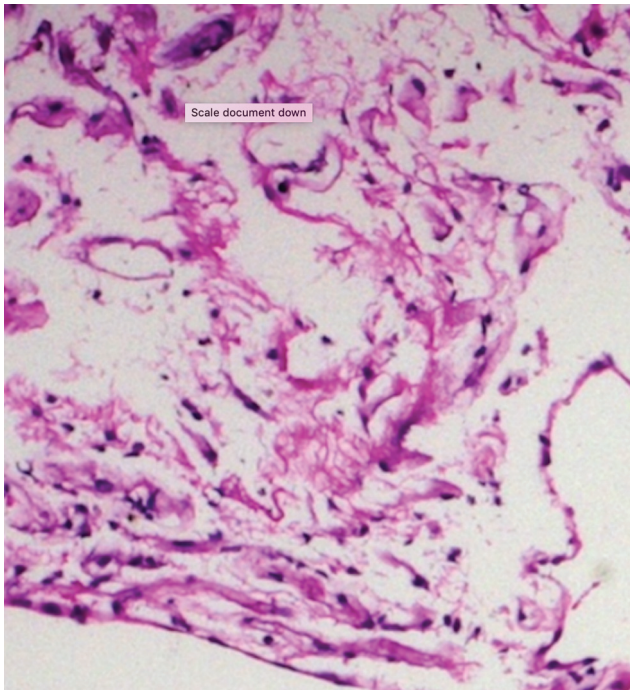

Fig. 2 Histopathological examination of the lesion (hematoxylin and eosin stain) showing a thin cyst wall formed of delicate fibrous connective tissue lined by meningothelial cells.

With the widespread availability of MRI, more and more intramedullary arachnoid cysts are being diagnosed. Asymptomatic cysts require proper counseling about possible natural evolution, while symptomatic cysts should be promptly treated. Though the best surgical technique is debatable, the outcome of postoperative neurological recovery is invariably good.

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Conflict of Interest
None declared.

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