

Case Report

# Large intradural craniospinal arachnoid cyst: A case report and review of literature

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## Abstract

Presence of an arachnoid cyst at craniospinal junction is not very common. This is a very rare anatomic site, with only seven other cases reported in the literature. We report a case of large intradural craniospinal arachnoid cyst presenting with obstructive hydrocephalus and cranial nerve palsy. A 39-year-old male presented with 8-month history of neck pain, headache, vomiting, visual disturbances, diminished taste sensation, and numbness of face. He had bilateral papilledema on ophthalmoscopy. Magnetic resonance imaging (MRI) revealed a posterior fossa arachnoid cyst extending down to the lower border of C5 vertebra. Posterior decompression was done through C5 laminectomy. He made a full recovery and was asymptomatic at 6-month follow-up examination. The clinical features, diagnosis, and management of these rare craniospinal arachnoid cysts are discussed.

**Key words:** Cranial nerve palsy, craniospinal junction, hydrocephalus, large arachnoid cyst

## INTRODUCTION

Arachnoid cysts are benign intra-arachnoid collections of cerebrospinal fluid and comprise around 1% of the intracranial masses. Arachnoid cysts of the craniospinal junction are very rare lesions,<sup>[1]</sup> with only seven cases reported in the literature. But any case presenting with such a large craniospinal arachnoid cyst, as it is in ours, has yet to be reported. They produce a range of clinical presentations resulting from hydrocephalus and compression of the underlying structures.<sup>[2]</sup> Interestingly, no features of cerebellar or spinal compression were found in our case. The size, location of the cyst, and clinical presentation were the uncommon features of this case.

## CASE REPORT

A 39-year-old male presented with 8-month history of progressively increasing neck pain and headache. To start with, the headache was suboccipital, which gradually became holocranial. He also complained of worsening vomiting over previous 3 months. For the past 20 days, he had developed numbness of face, diminution of vision in both eyes, and diminished taste sensation in the anterior part of tongue. No other complaint was reported. There was no past history of trauma to the craniospinal region.

On neurological examination, visual acuity was found to be 6/36 bilaterally, with fundus showing papilledema. There was diminution of corneal reflex, with decreased sensation over V1, V2, and V3 divisions. Taste sensation from anterior two-thirds of the tongue was diminished. No other neurological abnormality was found. All routine laboratory investigations were within normal range. No congenital malformations were present.

Plain X-ray of the cervical spine was normal. Computed tomographic (CT) scan of head showed a well-circumscribed extra-axial midline hypodense lesion in the posterior fossa [Figure 1]. Magnetic resonance imaging (MRI) brain revealed posterior fossa midline extra-axial well-margined

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T1WI hypointense, T2WI hyperintense non-enhancing cystic lesion with elliptical elongation below the foramen magnum [Figure 2a, b, and d]. There was associated mass effect compressing the 4th ventricle, causing non-communicating hydrocephalus [Figure 2c]. MRI of craniocervical axis revealed the same cystic lesion extending down through foramen magnum up to the lower border of C5 vertebra, communicating with the spinal subarachnoid space, compressing and displacing the medulla oblongata and proximal cervical cord [Figure 3].

The patient underwent posterior decompression through C5 laminectomy. Upon opening the dura, a bluish thin-walled cyst was encountered dorsal to the spinal cord. Clear fluid was aspirated from the cyst and a partial excision of the cyst wall was carried out. Histopathologic examination of the cyst wall confirmed it to be an arachnoid cyst [Figure 4]. The patient rapidly improved following the surgery. CT scan done on 8th postoperative day revealed decreased size of the lesion [Figure 5]. He made a full recovery and was asymptomatic at 6-month follow-up examination.

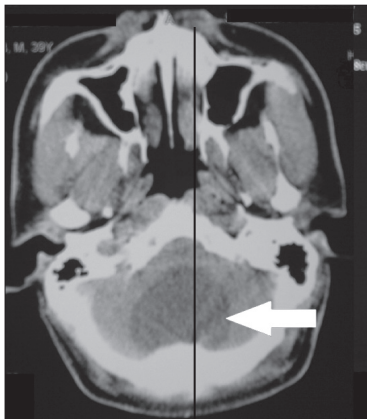


Figure 1: CT scan brain axial image showing well-circumscribed extra-axial midline hypodense lesion in the posterior fossa (arrow)

## DISCUSSION

The posterior fossa is the second most common location of arachnoid cysts, after the middle cranial fossa.<sup>[3-5]</sup> Symptomatic arachnoid cysts of the craniocervical junction are rare, with only seven cases reported in the literature, the largest one extending up to the lower level of C4 vertebra.<sup>[6]</sup> In our case, the cyst extended up to the lower level of C5 vertebra.

Most of these cysts are congenital. Consequently, about 75% of those causing symptoms are diagnosed in young children. It is not known how exactly arachnoid cysts grow, but when they give rise to symptoms, these are usually caused by local compression of the surrounding nervous structures and/or obstructive hydrocephalus.<sup>[7]</sup> Ventral arachnoid cysts at the craniocervical junction present because of compression of corticospinal tracts.<sup>[6]</sup> Our case had no sign and symptoms of corticospinal tracts' compression. Probably it was due to the location of the

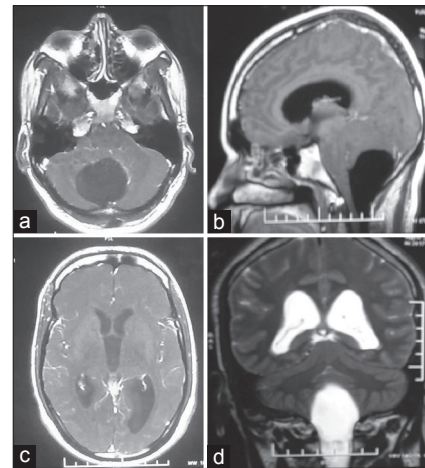


Figure 2: MRI brain: (a) Axial and (b) Sagittal views showing posterior fossa extra-axial well-margined T1-hypointense, (c) T2-hyperintense non-enhancing midline cyst with hydrocephalus. (d) Coronal view shows elongation of the cyst below foramen magnum

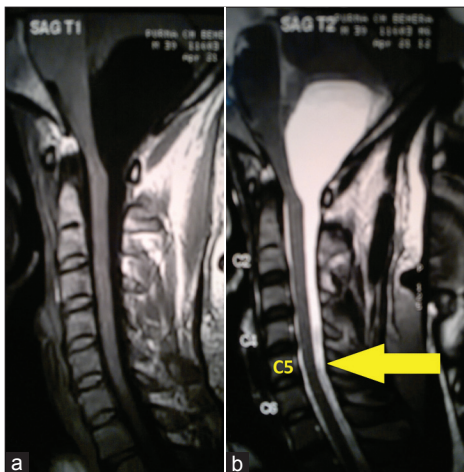


Figure 3: MRI craniocervical axis sagittal view showing posterior fossa T1WI hypointense, (a) T2WI hyperintense. (b) Cystic mass communicating with the spinal subarachnoid space, compressing and anteriorly displacing the medulla oblongata and proximal cervical cord up to the lower border of C5 (arrow)

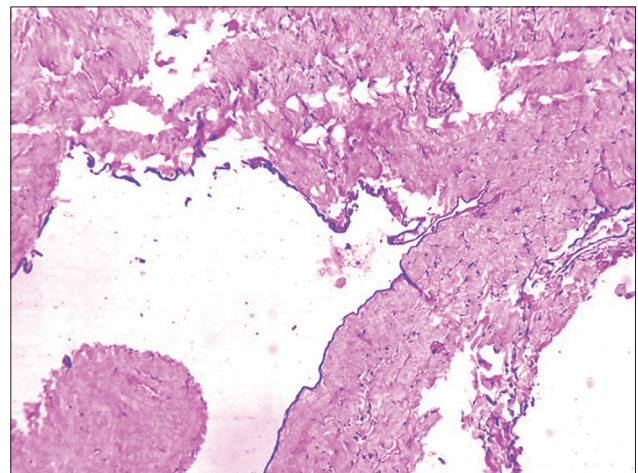


Figure 4: Photomicrograph of the cyst wall demonstrating a thin arachnoid layer lined by discrete nests of meningotheelial cells



Figure 5: Postoperative decreased size of the cyst

cyst dorsal to the medulla, which spared the more ventrally placed corticospinal tracts. Compressing the trigeminal sensory nucleus and nucleus tractus solitarius with relative sparing of cerebellum explains the signs and symptoms noted in our case.

Galassi *et al.*<sup>[3]</sup> suggested that the clinical picture is one of long duration in adults. Some complaints, such as headache, can have a long history reflecting a chronic increased intracranial pressure, but the duration of signs and symptoms, such as cranial nerve palsy, was usually short as it is in our case. Similar to the report published by Erdinçler *et al.*,<sup>[8]</sup> our case showed a decrease in ventricular size following simple partial cyst excision. Thus, it seems mechanical obstruction of the CSF circulation pathways by the cyst may be the main cause of hydrocephalus.

Imaging findings of all craniospinal arachnoid cysts reveal a large posterior fossa arachnoid cyst extending through the foramen magnum to the level of the upper spine. Differentiation of the posterior fossa arachnoid cysts from other cystic lesions is essential for proper treatment planning. The characteristic appearance of an uncalcified, low-density, extra-axial mass with regular borders that do not enhance with administration of contrast medium makes the differential diagnosis from neoplastic or infectious disorders simple in most cases.

The available therapeutic alternatives range from total extirpation of the cyst to a cystoperitoneal shunting procedure.<sup>[9-11]</sup> Total excision may not always be feasible owing to the presence of multiple adhesions between the cyst and the surrounding structures. Incision and deroofting of the cyst is, therefore, the more commonly performed procedure.<sup>[12]</sup> Our patient was managed by partial cyst

Panigrahi, *et al.*: Large intradural craniospinal arachnoid cyst wall excision at its lower<sup>[13]</sup> end via C5 laminectomy, resulting in adequate decompression and rapid clinical improvement.

## CONCLUSION

Features of cerebellar and spinal compression may not be present even in large craniospinal arachnoid cysts. Mechanical obstruction at the level of 4<sup>th</sup> ventricle or aqueduct of Sylvius is the major underlying cause of hydrocephalus in adult patients with craniospinal arachnoid cysts, and this is preventable with the restoration of CSF circulation pathways by cyst excision without the need for shunting. Cyst wall excision at the lower end via a small laminectomy is equally effective and less invasive.

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