

# Anterior cervical meningocele with craniovertebral junction instability – A case report and literature review

### ABSTRACT

Anterior meningocele involves herniation of meninges through an abnormal defect in the anterior vertebral column. The pathogenesis, natural history, and management strategy of anterior cervical meningocele (ACM) are uncertain. We report a case of ACM with high cervical instability in a case of neurofibromatosis 1. Unlike other reported cases, torticollis and instability due to ACM were the major concerns in this case. We aim to discuss the management strategy and surgical nuances of such cases.

**Keywords:** Anterior cervical meningocele, craniovertebral junction instability, dural ectasia, mesodermal dysplasia, neurofibromatosis type 1

### INTRODUCTION

An anterior meningocele involves herniation of meninges through an abnormal defect in the anterior vertebral column while neural elements remain intact. Anterior cervical meningocele (ACM) affecting craniovertebral junction has never been reported previously. Its occurrence in the high cervical region posed torticollis and craniovertebral junction instability as the major concerns. We aim to discuss the management strategy and surgical nuances of such cases.

### CASE REPORT

A 19-year-old female had presented with torticollis since childhood, progressive vision loss in both eyes for 4 months, and cerebellar symptoms for 3 months. The fundus examination study was suggestive of early papilledema. Bilateral cerebellar signs were positive. There were multiple café au lait spots and freckles over the trunk. There were no symptoms or signs attributable to compression by the sac.

### Imaging

The magnetic resonance imaging (MRI) brain revealed thickened bilateral optic nerves and left cerebellar pilocytic

astrocytoma with proximal hydrocephalus [Figure 1]. The meninges were seen herniating anteriorly into the retropharyngeal space at C2, C3, and C4 vertebral level. No neural elements were seen within the sac [Figure 2a and b]. On computed tomography (CT) cervical spine with three-dimensional reconstruction, the right lateral mass and part of the body of C3 and C4 vertebrae were absent. The right C2 pars and pedicle were also deficient [Figure 3a and b]. The posterior elements of C2 and C3 were fused. Dynamic X-rays of the cervical spine revealed abnormal mobility at the C3-C4 level along with atlanto-axial dislocation [Figure 3c and d].

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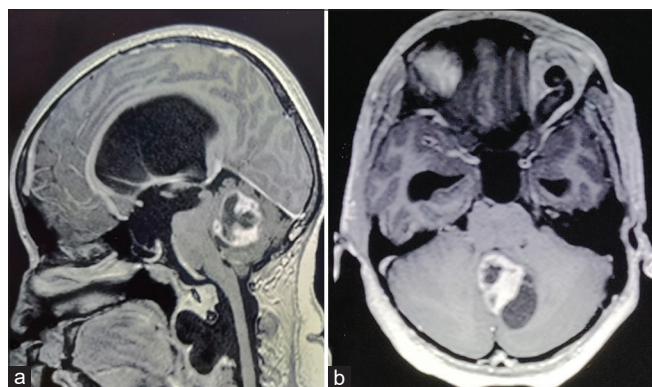
## Management

Under general anesthesia, intubation with a laryngoscope was performed. No compression due to the ACM was seen that could make intubation difficult. Care was taken to completely immobilize the neck while the patient was turned to the prone position. The head was stabilized in the sugita frame in a neutral position. Following midline

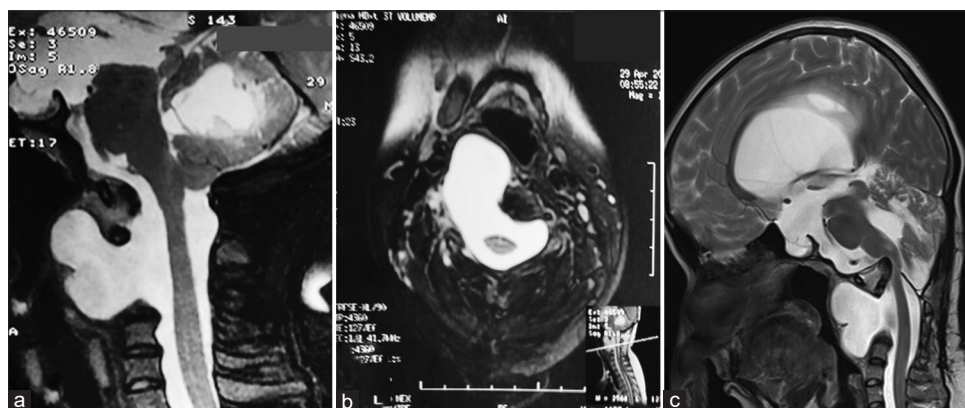
suboccipital craniectomy, the dura was seen bulging between the C1 and the C2 posterior elements. The sac reduced in size on opening the dura. Following tumor excision, the C1-C2 joint was exposed. The right C2 nerve root was cut that showed egress of cerebrospinal fluid (CSF), causing further decompression of the thecal sac. The right C2 pars and pedicle were hypoplastic with a tilt to the right side. Bilateral C1, C3, and C4 lateral mass screws were placed and fixed with a rod. An external lumbar drain was placed intraoperatively as a preventive measure against postoperative CSF leak.

## Postoperative recovery

The postoperative scan showed total excision of the tumor with correction of torticollis. She was planned for chemotherapy for optic nerve glioma. The external lumbar drain was removed on postoperative day 5. Figure 4 shows postoperative CT cervical spine showing the fixation. A follow-up MRI cervical spine was done in the 18<sup>th</sup> month. The anterior meningocele was of the same size as in the preoperative scan [Figure 2c]. The patient continues to be asymptomatic for ACM at 18 months of follow-up.



**Figure 1:** (a) Magnetic resonance imaging (head) – T1 contrast sagittal section and (b) T1 contrast axial section, showing left cerebellar pilocytic astrocytoma. The tumor is compressing the fourth ventricle and the aqueduct causing marked distension of the third and lateral ventricles. The anterior cervical meningocele is seen in the sagittal section



**Figure 2:** (a) Preoperative magnetic resonance imaging cervical spine – T2 sagittal and (b) T2 axial images showing anterior cervical meningocele. It is compressing the oropharynx anteriorly. There are no neural elements seen within the sac. The postoperative magnetic resonance imaging sagittal section (c) showed no change in the size of the anterior cervical meningocele. There is no residual tumor. Postoperative changes are noted in the cerebellum

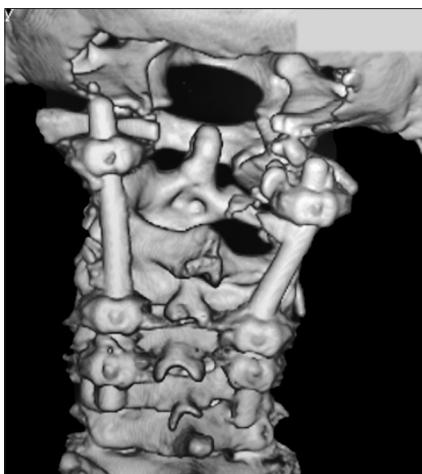


**Figure 3:** (a and b) Computed tomography cervical spine three-dimensional reconstruction showing torticollis toward the right side. C2 and C3 are fused. The inferior part of the right pedicle and pars of C2 and the right pedicle of C3 and C4 are absent. (c and d) X-ray cervical spine – neutral and flexion showing instability at C1C2 and C3C4 levels in the form of increased atlantodental interval and anterolisthesis, respectively

**Table 1: Literature review of reported cases of anterior cervical meningocele**

Author	Age/ gender	Level	Presenting complaints	Compressive Symptoms due to sac	Associated syndrome	Management	Change in size of the sac on follow-up (months)	Outcome/follow-up (months)
Shore <i>et al.</i> , 1982 <sup>(1)</sup>	New born/ female	C2-C3	Torticollis	Neck mass	No neuro-cutaneous markers	Cyst excision done with d/d of cystic hygroma	NA	Postoperative CSF leak and meningitis- managed with antibiotics
O' Neil <i>et al.</i> , 1983 <sup>(2)</sup>	51/female	C4-5	Neck pain on movement (mild)	Nil	NF1	NA	NA	NA
Kaiser <i>et al.</i> , 1986 <sup>(3)</sup>	40/male	C3	Neck pain Right upper limb weakness	Dysphagia	NF1	Conservative	NA	NA
So and Li 1989 <sup>(4)</sup>	55/female	C3-T2	Low backache	Nil	NF1	Conservative	NA	NA
Freund and Timon 1992 <sup>(5)</sup>	59/female	C3-C5	Neck pain Right shoulder pain	Right side neck Swelling Dysphagia Change in voice	NF1	NA	NA	NA
Göçer <i>et al.</i> , 1999 <sup>(6)</sup>	44/male	C5-7	Neck pain radiating to left arm Weakness of intrinsic muscles of left hand	Nil	NF1	Left C5 hemilaminectomy with excision of the C5-6 neuroma Attempted repair of sac, but failed	No change (6 months)	Clinically stable (6 months)
Kos <i>et al.</i> , 2009 <sup>(7)</sup>	49/male	C3-6	Nil	Dysphagia, Dysphonia, Obstructive sleep apnea syndrome	NF1	Tracheostomy and excision (anterior approach)	NA	Postoperative CSF leak- managed with external lumbar drain
Gallagher <i>et al.</i> , 2015 <sup>(8)</sup>	45/female	C6-7	Neck pain (following fall)	Nil	KFS	Conservative	No change (6 month)	Asymptomatic (6 months)
Present case	19/female	C2-C4	Torticollis Neck pain Raised ICP headache (Cerebellar pilocytic astrocytoma) Diminution of vision (optic nerve glioma - B/L)	Nil	NF1	C1C3C4 posterior screw and rod fixation, excision of cerebellar pilocytic astrocytoma with lax duraplasty (artificial dura)	No change (6 month)	Asymptomatic (18 months)

NF1 - Neurofibromatosis type 1; KFS - Klippel-Feil syndrome; NA - Not applicable; CSF- Cerebrospinal fluid , ICP- Intracranial pressure; B/L- Bilateral



**Figure 4: Post-operative CT cervical spine 3D reconstruction with instruments in situ (posterior view). Craniectomy defect and the cervical vertebral anomalies are also appreciated**

## DISCUSSION

Anterior meningocele is most commonly seen in the sacral region, followed by thoracic and lower cervical levels. To the best of our knowledge, eight cases of anterior or anterolateral cervical meningocele have been reported. This is the second case where C2 was also involved, leading to torticollis and high cervical instability. The pathogenesis, natural history, and management strategy of ACM are uncertain. Table 1 summarizes the clinical presentation, management, and outcome in the reported cases.

Neurofibromatosis is associated with mesodermal dysplasia and dural ectasia. The mesodermal dysplasia in NF1 causes vertebral body defect with subsequent anterior herniation of the dysplastic meninges leading to anterior meningocele. We concur with this most acceptable hypothesis. It explains the occurrence of the large bony defect involving three levels and the vertebral bodies seen in this young patient. Furthermore, neurofibromatosis 1 was the underlying syndrome in seven out of the nine cases of spontaneous ACM.<sup>[1-8]</sup> Another hypothesis is the progressive erosion of widened intervertebral foramina in neurofibromatosis. This is supported by the findings of the authors who reported multiple anterior and anterolateral meningoceles. Other possibilities such as cystic degeneration of neurofibroma, trauma, or elongated nerve sheaths are less likely. ACM is nondysraphic as against the posterior cervical meningoceles.

Only two cases, including the present case, had ACM involving CVJ. Both these patients were young and had torticollis since birth. All other patients were having subaxial cervical spine

ACM and their average age was 49 years (range 40–59 years). Neck pain was the most common presenting complaint. Airway compromise and dysphagia as a result of mass effect could be a major complication due to the sac. Despite a large sac, our patient had none of these symptoms. The intubation was smooth with no airway compromise. Only three out of nine cases of cervical meningocele had presented with dysphagia and dysphonia.<sup>[3,5,7]</sup> In all other cases, ACM was an incidental finding.

There are two aspects of ACM management-ACM sac repair and instability due to bony defect. Despite the theoretical possibility of ACM causing immense morbidity by its location, it remains essentially benign. Only one in nine patients needed intervention for ACM sac for severe compressive symptoms.<sup>[7]</sup> Similar to this case, two other authors reported nonprogression of sac size in the follow-up. It is to be noted that there is no neural component in the ACM (unlike myelomeningocele) or any associated neurological deficits due to the sac. In this given scenario, any attempt to repair it at this site would do more harm than good. Insertion of lumbo-peritoneal shunt has been reported to allow decompression of anterior meningocele. As there were no symptoms and no progression of the sac, we preferred to avoid a hardware placement. The cervical spine instability due to bony defect must be addressed. Due care should be taken during intubation and positioning of these patients.

## CONCLUSION

Congenital ACM is pathognomic of NF1, occurring as a consequence of mesodermal dysplasia and dural ectasia. It is a rare cause of craniovertebral junction instability. Given a benign course, management is conservative. Surgical intervention is indicated for the secondary instability or rarely for compressive symptoms due to the sac.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

## REFERENCES

1. Shore RM, Chun RW, Strother CM. Lateral cervical meningocele. *Clin Pediatr (Phila)* 1982;21:430-3.
2. O'Neill P, Whatmore WJ, Booth AE. Spinal meningoceles in association with neurofibromatosis. *Neurosurgery* 1983;13:82-4.
3. Kaiser MC, De Slegte RG, Crezée FC, Valk J. Anterior cervical meningoceles in neurofibromatosis. *AJNR Am J Neuroradiol* 1986;7:1105.
4. So CB, Li DK. Anterolateral cervical meningocele in association with neurofibromatosis: MR and CT studies. *J Comput Assist Tomogr* 1989;13:692-5.
5. Freund B, Timon C. Cervical meningocele presenting as a neck mass in a patient with neurofibromatosis 1. *J Laryngol Otol* 1992;106:463-4.
6. Göçer AI, Tuna M, Gezercan Y, Boyar B, Bağdatoğlu H. Multiple anterolateral cervical meningoceles associated with neurofibromatosis. *Neurosurg Rev* 1999;22:124-6.
7. Kos MP, Peerdeman SM, David EF, Mahieu HF. Multiple cervical anterior meningoceles in a patient with neurofibromatosis type 1 cause dysphagia and dyspnoea. *Otolaryngol Head Neck Surg* 2009;140:612-3.
8. Gallagher MJ, Chavredakis E, Carter D, Bhojak M, Jenkinson MD, Clark SR. Cervical vertebral fusion with anterior meningocele. *Neuroradiol J* 2015;28:205-8.