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Case Report

Neurenteric cyst of the dorsal craniocervical junction in an adult: A case report and operative video

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ABSTRACT

Background: Neurenteric cysts (NCs) are rarely located in the dorsal craniocervical junction (CCJ).

Case Description: Here, we present a case and show the surgical video of a 24-year-old man with a history of neck pain, progressive hemiparesis, and difficulty swallowing. Radiological images revealed an intradural extramedullary lesion at the dorsal CCJ. Posterior approach with C1–C2 laminectomy for resection of the lesion was performed with significant improvement in symptoms postoperatively. Histopathological examination confirmed the diagnosis of NC.

Conclusion: This case demonstrates a rare location of a NC in an adult patient. Complete excision of the cyst wall and its content is the recommended treatment option.

Keywords: Craniocervical junction, Endodermal cyst, Foramen magnum, Neurenteric cyst

INTRODUCTION

Neurenteric cysts (NCs) (also known as endodermal, enterogenous, and bronchogenic cysts) are benign, rare, congenital lesions that are lined by endodermal cell-derived epithelium.[1,13] These cysts are thought to arise during early embryogenesis from abnormal communication between the endodermal and ectodermal tissues.[3,11] NCs can be located anywhere along the neuroaxis, and in most cases, they are located in the lower cervical and upper thoracic spine and are typically ventrally located.[11] NCs are rarely located in the craniocervical junction (CCJ). Moreover, dorsally located CCJ NCs are extremely rare as only five similar cases have been reported in the adult population. Herein, we describe a case of NC of the dorsal CCJ in an adult patient along with a surgical video.

CLINICAL PRESENTATION

History and examination

A 24-year-old man presented with a 4-week-history of neck pain, progressive hemiparesis, and mild dysphagia. Neurological examination revealed an increase in tone in the right upper and

lower extremities, a power of 4/5 in the proximal muscle group, and a power of 3/5 in the distal muscle groups of the right upper extremities. He also had a power of 4/5 in the right lower extremity. On the left side, he had a normal tone in both upper and lower limbs and a power of 4/5 in both upper and lower extremities. He had positive Hoffman sign and brisk reflexes bilaterally. Cranial nerves examination revealed weakness in the right accessory nerve with a power of 4/5 for shoulder elevation; however, the remaining cranial nerves were intact.

Imaging studies

A plain head computed tomography scan and upper cervical spine revealed a hypodense craniocervical lesion up to the C2 level. Magnetic resonance imaging (MRI) of the cervical spine showed an intradural extramedullary lesion centered in the dorsal CCJ measuring 2.2 × 1.9 × 2.6 cm on AP, transverse, and CC dimensions, respectively. It showed increased signal intensity and a fluid-fluid level on T2 sequence and low signal intensity on T1 sequence. The lesion did not show enhancement with gadolinium, with no suppression of fluid-attenuated inversion-recovery sequences and no diffusion restriction on diffusion-weighted imaging [Figure 1].

Operation

With the aid of intraoperative neuromonitoring, a suboccipital craniotomy with C1–C2 laminectomy was performed to reach the lesion. The patient was placed in the prone position with the head fixed in three-point fixation device. A midline skin incision was made started from the inion to C2. Suboccipital craniotomy was performed followed by C1–C2 laminectomy. The dura was opened starting caudally and proceeding cranially. Tuck-up dural sutures were applied. The lesion came into view; and it was observed to be round, yellowish, with a pedicle attached to the dorsal cord, and traversed with multiple C1 rootlets; subsequently, the arachnoid was carefully incised. Arachnoid attachments were dissected, and the lesion was progressively mobilized. The central mass of the cyst was aspirated and debulked using suction in an attempt to create a safe plane for dissection. The wall of the cyst was dissected and elevated from the obex and the surrounding nerve roots and was ultimately resected. The dura was closed in a watertight fashion using standard measures [Video 1].

Postoperative course and follow-up

The subsequent histopathological examination confirmed the diagnosis of NC [Figure 2]. Postoperatively, the patient showed complete resolution of his weakness. Follow-up MRI showed gross-total resection [Figure 3].

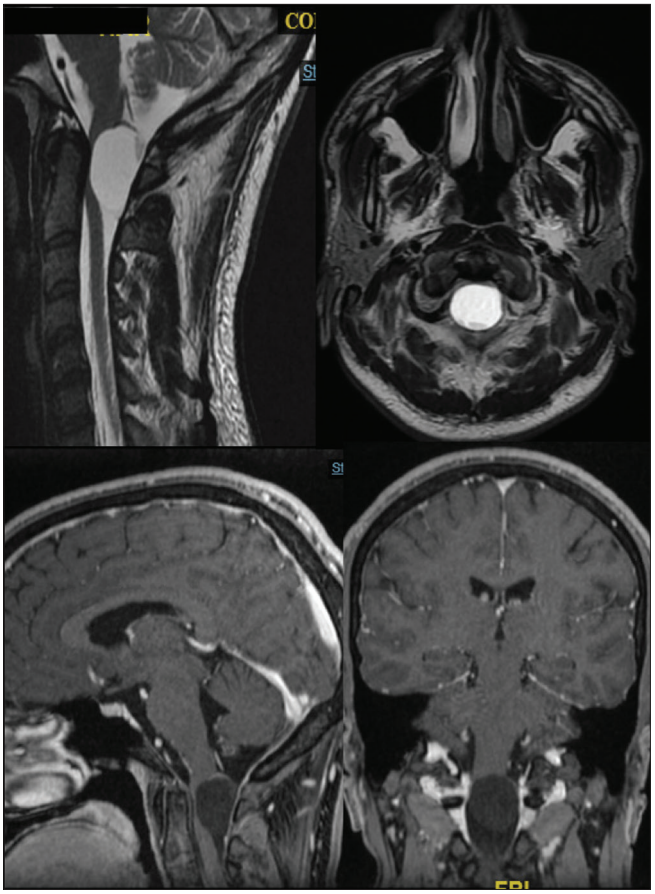
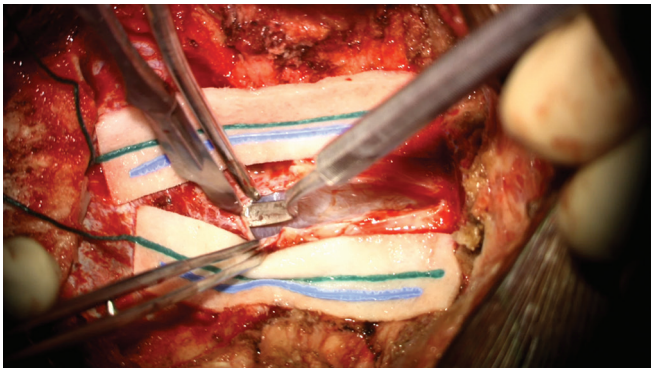


Figure 1: Preoperative imaging. Magnetic resonance imaging of the cervical spine showing an intradural extramedullary lesion centered in the dorsal craniocervical junction measuring 2.2 × 1.9 × 2.6 cm on AP, transverse, and CC dimensions, respectively. It showed increased signal intensity and a fluid-fluid level on T2 sequence and low signal intensity on T1 sequence.



Video 1: Operative video.

DISCUSSION

NCs are rare congenital lesions that represent 0.3–1.3% of all spinal lesions.[3,16] They are typically located in the lower cervical and upper thoracic spine with rare instances of cysts

located in the CCJ,[9,11,14] cerebellopontine angle,[4,15] and supratentorial compartments.[5,7] In this article, we describe a case of NC of the dorsal CCJ in an adult. NCs are typically located ventrally and have been associated with other congenital bony anomalies, such as scoliosis, fusion, or failed segmentation of multiple vertebrae associated with Klippel–Feil syndrome, hemivertebrae, and spina bifida. Ohba *et al*.[14] reviewed 36 cases of CCJ NCs but none were located dorsally. A careful review of the literature identified five cases of dorsally located CCJ NCs in adults [Table 1].

The embryogenesis of NCs remains to be elucidated. Proposed theories include persistence of the neurenteric canal, abnormal vascular supply to the developing neural tissue, or split notochord syndrome.[4,8,10]

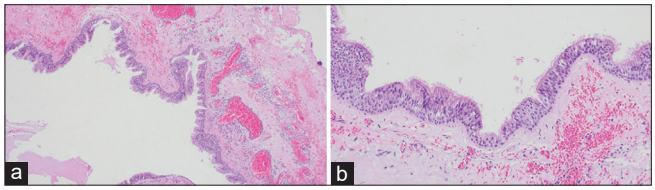


Figure 2: Histopathological imaging. (a) Cyst wall lined by pseudostratified columnar epithelium with ciliated cells, resting on fibrovascular layer. (b) Higher power shows cyst wall lined by pseudostratified columnar epithelium with cilia. Goblet cells are present.

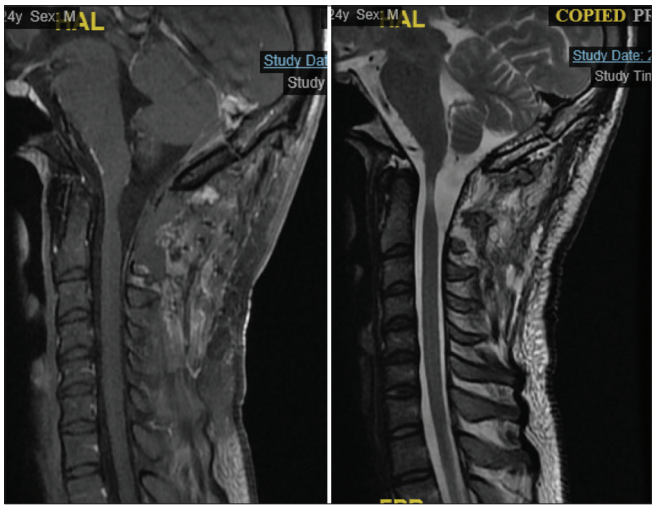


Figure 3: Postoperative imaging. MRI of the cervical spine showing complete resection of the lesion.

Table 1: Summary of reported cases of neurenteric cysts of the dorsal craniocervical junction in adults.

Author	Age/gender	Presentation
Bolcha *et al*., 2012	27/F	Lower cranial nerve symptoms and lower limbs sensory loss
Clare *et al*., 2006	57/F	Neck pain
Filho *et al*., 2001	30/F	Ataxia, right hand numbness
Gu *et al*., 2005	39/F	Headache, vertigo, ataxia
Weiss *et al*., 1996	32/M	Meningismus, headache

NCs have a male predilection and are commonly documented during the 2nd or 3rd decade.[6,12] These cysts can cause symptoms by inflammatory reactions secondary to leakage of the contents of the cyst or mass effect due to cyst expansion. Common presentations of CCJ NCs include neck pain, myelopathy, radiculopathy, and lower cranial nerve symptoms. On MRI, these lesions show features relevant to their protein and mucinous content including high signal intensity on T2 and low or isointensity on T1 sequences. The peripheral enhancement of the cyst wall has been demonstrated in a few cases; however, they generally lack avid enhancement.[2,15]

Although few cases have shown spontaneous regression,[9] many neurosurgeons advocate complete excision of the cyst wall and its content, if feasible as the main treatment strategy. However, in cases, where complete resection cannot be achieved without risking associated critical structure, subtotal resection with careful follow-up is a reasonable option. The risk of recurrence reaches up to 37% regardless of location in one series.[11] Simple aspiration of the cyst is not recommended as it has been associated with higher risk of recurrence.[11]

The appropriate surgical approach for CCJ NCs depends largely on the location of the cyst. Earlier reports have shown the feasibility of anterior transoral approach and far lateral transcondylar approach for ventrally located cysts. For dorsally located cysts, the posterior approach is advocated as it provides adequate results. It allows early aspiration and decompression of the cyst before manipulating the cyst capsule; therefore, it reduces the risk of injury to neighboring critical structures. Patients with a remnant cyst wall require careful follow-up with serial imaging to detect recurrence.

CONCLUSION

Dorsally located CCJ NCs are rare. Complete resection of the cyst wall and its contents is of paramount importance to prevent recurrence.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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