CASE REPORT

Neurenteric cyst at the craniovertebral junction: A report of two cases

Sathwik Raviraj Shetty, Manas Panigrahi, Satish Rao[1]

Departments of Neurosurgery, and [1]Pathology, Krishna Institute of Medical Sciences, 1-8-31/1, Minister Road, Secunderabad, Andhra Pradesh, India

ABSTRACT

Neurenteric cysts at the craniovertebral junction are extremely rare, in that most are located ventrally in the midline. We report two patients with neurenteric cysts in an unusual location – the lateral craniovertebral junction. The embryological basis, clinical features, imaging characteristics, and management options in these patients have been discussed.

Key words: Cervicomedullary junction, craniovertebral junction, enterogenous cyst, far-lateral approach, neurenteric cyst

Introduction

Neurenteric cysts (NCs) or enterogenous cysts are rare benign cysts that are lined by the endodermal-derived epithelium. NCs of the central nervous system are most commonly seen in the lower cervical and upper thoracic region, making up to 0.3-1.3% of all spinal cord tumors.[1] Intracranial NCs are extremely rare. Only about 80 cases have been reported in the literature, most occurring in the midline posterior fossa, occasional in the cerebellopontine angle and fourth ventricle, and very rarely in the cerebral hemispheres.[2-4]

The NCs at the craniovertebral junction (CVJ) are even rarer; fewer than 40 cases have been reported in literature till date.[5-7] We describe two patients who were treated surgically for a symptomatic NC at the CVJ and discuss their diagnostic and therapeutic considerations.

Case Reports

Case 1

A 7-year-old girl presented with complaints of gradually progressing neck pain of 7 months duration. The pain was localized in the neck and aggravated with neck movements. On examination, she had no focal neurological deficits and her laboratory investigations were normal. Magnetic resonance imaging (MRI) done revealed an intradural extramedullary lesion extending from C1 to C4 region, located ventrolateral to the upper cervical cord [Figure 1]. The patient underwent C1, C2 hemilaminectomy and subtotal excision of the lesion. Postoperatively, the patient had relief from neck pain. The histopathologic examination (HPE) was suggestive of NC.

The patient was advised regular follow-up. However, the patient had recurrence of symptoms 6 months post-surgery and a repeat MRI done showed recurrence of the lesion. The patient was re-operated. This time, a right far-lateral approach was taken to facilitate access to the cyst located ventral to the upper cervical nerves. The cyst was decompressed and the cyst wall was completely excised [Figure 2]. Postoperative period was uneventful. The HPE showed a cyst lined by columnar mucinous epithelium [Figure 3].

Case 2

A 36-year-old lady presented with complaints of intermittent right-sided headache of 2 years duration with a 20-day history of diplopia, mild deviation of angle of the mouth to the left, and decreased hearing in the right ear. Neurological examination revealed right 6th, 7th, and 8th nerve paresis with cerebellar ataxia. Audiological investigation revealed a right sensorineural hearing loss. An MRI of the brain showed an extra-axial lesion in the right cerebellomedullary cistern, measuring 4 × 3 × 2.5 cm, extending from lower clivus up to the rim of foramen magnum [Figure 4]. The various possibilities considered were NC, epidermoid or dermoid cyst.

The patient was placed in the left lateral position, and a right far-lateral approach was taken. Intraoperatively, a thin-walled cyst was seen located ventral to the lower cranial nerves and extending from the 7th to the 8th complex up to the foramen

Access this article online	
Quick Response Code:	Website: www.asianjns.org
	DOI: 10.4103/1793-5482.125667

Address for correspondence:
Dr. Manas Panigrahi, Department of Neurosurgery, Krishna Institute of Medical Sciences, 1-8-31/1, Minister Road, Secunderabad, Andhra Pradesh, India. E-mail: manaspanigrahi@live.com

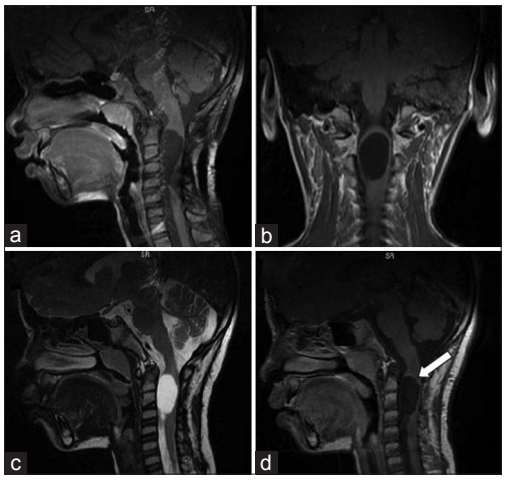

Figure 1: Preoperative images. (a and b) Sagittal and coronal T1W images showing a hypointense lesion extending from C1 to C4 region. (c) The lesion being hyperintense on T2W sagittal image. (d) Enhancing posterior wall on contrast (white arrow)

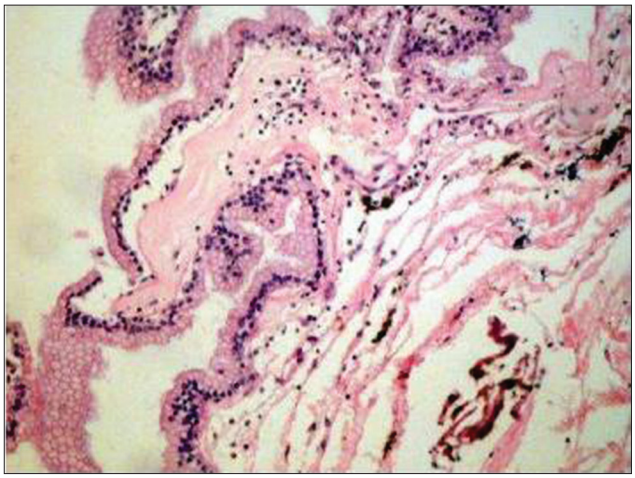

Figure 3: Cyst wall lined by columnar mucinous epithelium

magnum and extending into the jugular foramen. The cyst was decompressed by puncturing the cyst wall and aspirating its thick yellow contents. The cyst wall was gently separated and excised under Nerve Integrity Monitoring (NIM) [Figure 5]. The surgical cavity was irrigated with steroid impregnated saline. The postoperative period was uneventful.

The HPE was suggestive of NC with cyst wall lined with pseudostratified ciliated columnar epithelium [Figure 6].

Discussion

The NC is an unusual congenital cyst of endodermal origin, particularly rare at the CVJ.[5]

Embryology

NCs are thought to result from the disorder in the separation of the ectodermally derived spinal cord and the endodermally derived foregut during the closure of the neurenteric canal. Although this theory explains the appearance of spinal

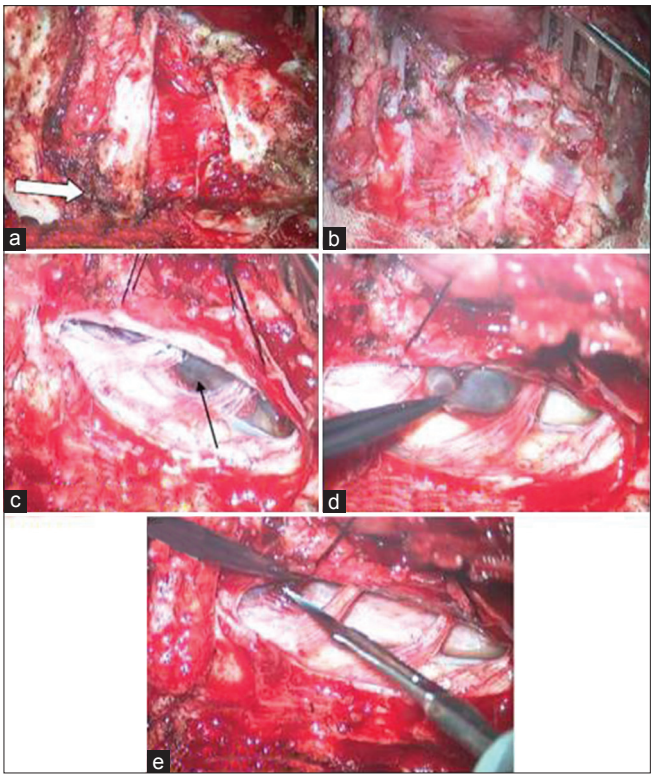

Figure 2: Intraoperative images. (a) Right far-lateral approach – the right C1 arch and right C2 arch exposed, (white) arrow indicating the midline. (b) Right posterior arch of C1 and right C2 lamina excised. (c) Cyst (black arrow) located ventral to upper cervical roots. (d) Excision of the cyst. (e) A remnant of the cyst wall being excised

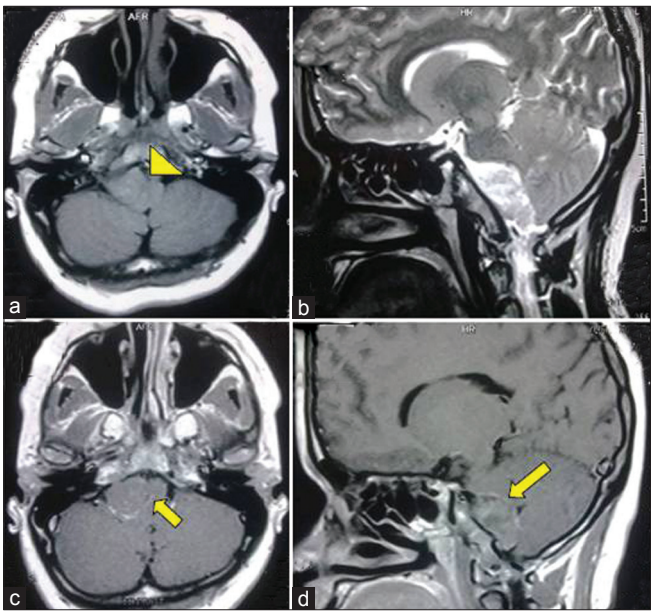

Figure 4: Preoperative images. (a) Axial T1W image showing an isointense lesion in the right cerebellomedullary cistern causing compression and distorting the lower medulla (arrow head). (b) Mixed density lesion on sagittal T2W images. (c and d) Contrast axial and sagittal images showing enhancement of the posterior wall (arrows)

NCs caudal to the clivus (which is the cranial margin of the endoderm in embryos) as in Case 1, it does not explain the

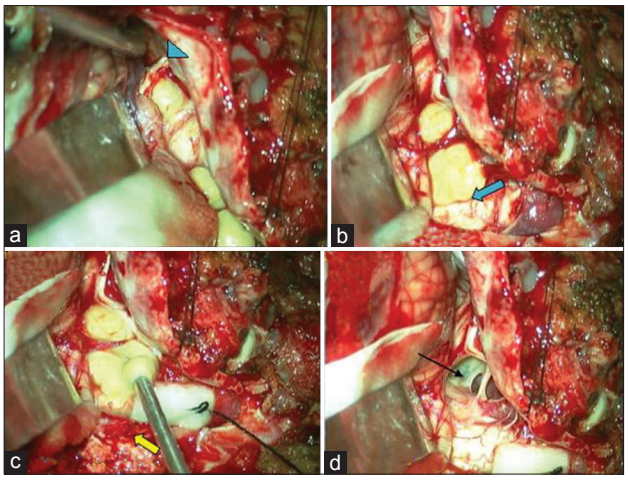

Figure 5: Intraoperative images showing the cystic lesion lying ventral to the cranial nerves. (a) Superior extent at the 7th–8th complex (blue arrow head). (b) Lower extent at the cervicomedullary junction (blue arrow). (c) Aspiration of the thick yellowish cystic material. (d) The cyst wall adherent to lower cranial nerves (black arrow) was excised

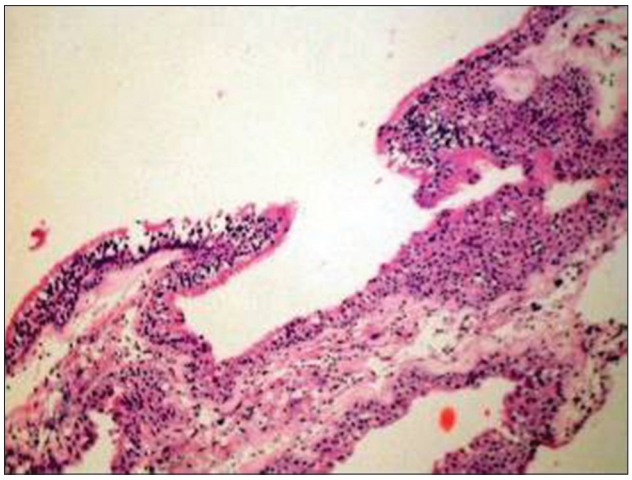

Figure 6: Cyst lined by pseudostratified ciliated columnar epithelium

presence of intracranial NCs or NCs rostral to the clivus as in Case 2. There are several theories as to the origins of intracranial NCs. Cheng *et al*.[8] suggested that they result from the rostral closure of the notochord by the mesenchyme forming the clivus. Graziani *et al*.[9] suggested that intracranial NCs are remnants of Seessel's pouch, a transient out-pouching of the oro-pharyngeal membrane at the cranial end of the foregut. This latter theory is supported by the fact that NCs share several immunohistochemical staining characteristics with Rathke's and colloid cyst. All three are positive for cytokeratin, epithelial membrane antigen (EMA), and carcinoembryonic antigen (CEA), and negative for glial fibrillary acidic protein (GFAP). NCs, in addition, are positive for CA19-9.[2] However, these theories do not explain laterally positioned intracranial NCs. Kulkarni *et al*.[10] put forth the theory of variable regression of endodermal–ectodermal adhesions at the cephalic end of the notochord which could explain the laterally positioned NCs rostral to the clivus.

Clinical features

Most CVJ NCs are located ventrally in the midline, anterior to the brainstem.[7] In our patients, the NCs were atypically located – laterally at the CVJ. Clinically, CVJ NCs can either mimic spinal NCs (relapsing and progressive myelopathy) or they can mimic intracranial NCs and present with symptoms from mass effect (headaches, cranial nerve involvement, gait disturbances, and long tract signs) or inflammation (recurrent aseptic meningitis). Both our patients presented with symptoms secondary to progressive direct mass effect.

Imaging

NCs can exhibit variable signal intensity characteristics depending upon the protein content of the cyst fluid. They can be either hypo, hyper, or isodense on the computed tomography (CT) scan. Most NCs are proteinaceous. On an MRI, they are iso to slightly hyperintense to cerebrospinal fluid (CSF) on T1W images and typically very hyperintense on T2W and FLAIR images. They may show a mild restriction on diffusion-weighted images with occasional cyst wall enhancement on contrast.[2] The imaging characteristics in our patients were typical of NCs except for the unusual location.

Unlike spinal NCs, intracranial and CVJ NCs are rarely associated with bony abnormalities.[2] Only two cases of CVJ NCs have been reported to have associated bony anomalies.[5] No associated bony anomalies were seen in our patients.

The differential diagnosis for CVJ NCs includes arachnoid cyst, epidermoid cyst, dermoid cyst, Rathke's cleft cyst, and schwannoma. Although a CT can exclude arachnoid and ependymal cysts (isodense to CSF), an MRI may be helpful in ruling out epidermoids (restricted diffusion), dermoid cysts (with fat suppression), and schwannomas (which enhance strongly). Only a pathologic examination using immunohistochemical staining can make a definitive diagnosis of NC.

Surgical approach

NCs are mostly located in front of the neural axis. The ideal approaches to NCs at the CVJ would be the transcondylar or a far-lateral approach.[6] Other approaches that have been employed are transoral, transclival endoscopic endonasal approach (EEA), suboccipital, retrosigmoid, and subtemporal approaches.[1] If an appropriate surgical approach is not employed, there is a high probability of subtotal resection and subsequent recurrence as seen in our patient (Case 1). By employing the far-lateral approach, we were able to achieve total excision in both our patients. For ventrally located NCs, the transcondylar approach might be ideal.

The best outcomes are associated with total removal of the cyst wall. The residual cyst wall is said to have proliferative potential and can also rarely undergo malignant transformation.[11,12] If the remnant walls are large enough to overlap, resealing of

the remnant cyst wall can occur with subsequent expansion of the cyst either by cyst epithelial secretion or by osmotic CSF accumulation. This was probably the cause of recurrence in our patient (Case 1). Hence, total excision should be the goal. However, if the cyst wall is densely adherent to the surrounding neurovascular structures, sub-total excision may be a reasonable option. During the operation, if small pieces of the remaining cyst wall can be distinguished from the arachnoid membrane and electrocoagulated to shrink them, overlapping can be avoided, thereby decreasing the chances of recurrence. Neither radiation nor chemotherapy has been recommended.[3]

Despite the apparent total cyst wall removal, a delayed recurrence is possible. Signs of aseptic meningitis or increasing levels of CA19-9 in the CSF are possible indicators of recurrence.[3,4] This necessitates extended serial follow-up imaging[4] for at least 10 years following surgery[13] and re-operation should be considered in cases of recurrence.

In conclusion, CVJ NCs are rare lesions often mimicking other cystic lesions on neuroimaging. The incidence of intracranial and CVJ NCs is gradually increasing. It is therefore important to be familiar with the clinical characteristics, imaging features, and management options of NCs.

References

1. Shi W, Cui DM, Shi JL, Gu ZK, Ju SQ, Chen J. Microsurgical excision of the craniocervical neurenteric cysts by the far-lateral transcondylar approach: Case report and review of the literature. Skull Base 2010;20:435-42.
2. Preece MT, Osborn AG, Chin SS, Smirniotopoulos JG. Intracranial neurenteric cysts: Imaging and pathology spectrum. AJNR Am J Neuroradiol 2006;27:1211-6.
3. Chaynes P, Bousquet P, Sol JC. Recurrent intracranial neurenteric cysts. Acta Neurochir (Wien) 1998;140:905-11.
4. Fujita T, Kayama T, Saito S, Yamakawa M, Nakai O. Immunohistochemical detection of tumor marker in recurrent clivus enterogenous cyst: Case report. Neurol Med Chir (Tokyo) 1997;37:479-82.
5. Abe K, Oyama K, Mori K, Ishimaru S, Eguchi M, Maeda M. Neurenteric cyst of the craniocervical junction-case report. Neurol Med Chir (Tokyo) 1999;39:875-80.
6. Liu JK, Couldwell WT. Far-lateral transcondylar approach: Surgical technique and its application in neurenteric cysts of the cervicomedullary junction. Report of two cases. Neurosurg Focus 2005;19:E9.
7. Ohba S, Akiyama T, Kanai R, Onozuka S, Kawase T. Endodermal cyst of the cranio cervical junction. Acta Neurochir (Wien) 2008;150:257-63.
8. Cheng JS, Cusick JF, Ho KC, Ulmer JL. Lateral supratentorial endodermal cyst: Case report and review of literature. Neurosurgery 2002;51:493-9.
9. Graziani N, Dufour H, Figarella-Branger D, Donnet A, Bouillot P, Grisoli F. Do the suprasellar neurenteric cyst, the Rathke cleft cyst and the colloid cyst constitute a same entity? Acta Neurochir (Wien) 1995;133:174-80.
10. Kulkarni V, Daniel RT, Haran RP. Extradural endodermal cyst of posterior fossa: Case report, review of the literature, and embryogenesis. Neurosurgery 2000;47:764-7.
11. Morita Y, Kinoshita K, Wakisaka S, Makihara S. Fine surface structure of an intraspinal neurenteric cyst: A scanning and transmission electron microscopy study. Neurosurgery 1990;27:829-33.
12. Sahara Y, Nagasaka T, Takayasu M, Takagi T, Hata N, Yoshida J. Recurrence of a neurenteric cyst with malignant transformation in the foramen magnum after total resection. Case report. J Neurosurg 2001;95:341-5.
13. Gauden AJ, Khurana VG, Tsui AE, Kaye AH. Intracranial neuroenteric cysts: A concise review including an illustrative patient. J Clin Neurosci 2012;19:352-9.

How to cite this article: Shetty SR, Panigrahi M, Rao S. Neurenteric cyst at the craniovertebral junction: A report of two cases. Asian J Neurosurg 2013;8:188-91.
Source of Support: Nil, **Conflict of Interest:** None declared.