



Neurenteric Cyst of the Craniovertebral Junction: Report of 2 Cases

Sathwik RS^{1,*}, Panigrahi M¹ and Satish Rao²

¹Department of Neurosciences, Krishna Institute of Medical Sciences, Minister Road, Secunderabad - 500003, AP, India

²Departments of Neurosurgery, Pathology, Krishna Institute of Medical Sciences, Minister Road, Secunderabad - 500003, AP, 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.

Keywords: neurenteric cyst; enterogenous cyst; craniovertebral junction; cervico-medullary junction; far lateral approach

***Corresponding author:** Dr. Sathwik R Shetty, Department of Neurosciences, Krishna Institute of Medical Sciences, Minister Road, Secunderabad - 500003, AP, India

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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 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 neurenteric cyst 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 localised in the neck and was aggravated with neck movements. On examination, she had no focal neurological deficits and her laboratory investigations were normal. The 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. Post operatively, the patient had relief from neck pain. The Histopathological 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). Post-operative 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 $4 \times 3 \times 2.5$ cms extending from lower clivus upto the rim of foramen magnum (Figure 4). The various possibilities considered were neurenteric cyst, 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-8th complex up to the foramen 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 NIM monitoring (Figure 5). The surgical cavity was irrigated with steroid impregnated saline. The post-operative period was uneventful.

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

Discussion

The Neurenteric cyst 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 NCs caudal to the clivus (which is the cranial margin of the endoderm in embryos) as in Case 1, it does not explain the 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-haryngeal 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, EMA, CEA and negative for 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 craniovertebral junction. 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

Neurenteric cysts 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 CT scan. Most NCs are proteinaceous. On an MRI they are iso to slightly hyperintense to 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 neurenteric cysts 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

Neurenteric cysts are mostly located in front of the neural axis. The ideal approaches to NCs at the CVJ would be the 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 reoperation 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 are gradually increasing. It is therefore important to be familiar with the clinical characteristics, imaging features and management options of NCs.

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