

Neurenteric cyst at the dorsal craniocervical junction in a child: case report

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Abstract

Neurenteric cysts, also known as enterogenous cysts, are uncommon, benign, congenital lesions that usually occur in the posterior mediastinum but can be seen at any level of the neuraxis. Here, we report a pediatric patient with a neurenteric cyst in the dorsal craniocervical junction as the only third reported pediatric case in the literature in this rare location, and describe the clinical course and pathologic findings with a review of the literature on this rare entity.

Keywords: dorsal, craniocervical, neurenteric cyst, pediatric

Introduction

Neurenteric cysts are uncommon, benign, congenital lesions that most commonly occur in the posterior mediastinum,[1] but can be encountered anywhere in the central nervous system (CNS) from the cranium to the coccyx.[2] The ventral spine is the most common location in the CNS.[3] Intracranial and craniocervical locations are even rarer and tend to occur in the posterior fossa, ventral to the brainstem.[4, 5] Here, we present the case of a neurenteric cyst in a 5-year old boy in a highly atypical location—the dorsal craniocervical junction extending from the inferior posterior fossa to the level of C3— and an atypical clinical course characterized by clinical fluctuation from spastic quadriplegia to completely normal neurologic examination, and review the literature on this rare entity.

Case report

History and examination

A 5-year-old boy presented to his local hospital in Haiti for evaluation of 2 years of progressive weakness. He had a normal prenatal period and birth and had developed normally until age 2, after which he developed slowly progressive right-sided weakness, followed by left lower extremity weakness, followed over the months prior to presentation by left arm weakness. His examination was notable for spastic paralysis of all limbs with the exception of the left upper extremity, with which he had minimal movement. Reflexes were symmetrically brisk with bilateral ankle clonus and Babinski signs. The neck was rigid, with severe pain on attempted passive neck movement. His cognition and cranial nerve examinations were normal. There was no prior history of trauma or infection, and no family history of neurologic disease with two healthy siblings. MRI revealed a dorsal cervicomedullary lesion extending to C3 with severe compression of the cervical spinal cord (Fig 1).

The decision was made to pursue charitable care in the United States, but this was delayed for almost a year due to administrative issues related to his visa. Over 3 months, the child recovered completely, and had a normal neurologic examination (including full strength, normal reflexes, and plantar flexor responses), with the exception of improved but persistent resistance to passive neck flexion, which was painful. Neuroimaging was repeated and unchanged.

Neuroimaging

MRI of the cervical spine demonstrated a 2.1 x 2.5 x 4.7 cm, well circumscribed, dorsal intradural, extramedullary cystic lesion extending from the level of the upper medulla to C3 with significant mass effect on the medulla and upper cervical cord. The lesion was T1 hypointense and T2 hyperintense with a thin peripheral capsule, and did not demonstrate diffusion restriction or enhancement on post-gadolinium sequences (Fig. 1). The lesion was slightly hypointense relative to CSF on MRI FIESTA, with no solid component, diffusion restriction or abnormal enhancement with gadolinium administration. There was no cord edema or associated syringomyelia, and the cerebellar tonsils were slightly superiorly displaced. The rest of the brain and cervical spine examination were normal, apart from expansion of the posterior elements at C1 and C2 to accommodate the cyst, indicating the long-standing nature of the cyst.

Operation

A suboccipital craniectomy with C1 and partial C2 laminectomy was performed for resection of the cystic lesion. Upon opening of dura, an intact thick-walled, yellow cyst was observed, with some calcifications near the wall at the foramen magnum. The cystic wall was adherent to the pia at the level of foramen magnum. The cyst and its contents were completely excised as confirmed by postoperative imaging (Fig. 2), and the dura was closed in a watertight fashion.

Postoperative course

The patient had an uneventful postoperative hospitalization and was discharged home on the 4th postoperative day with no neurologic deficit. He was seen back at one month, and was neurologically intact, now with normal range of motion of his neck. At 3 month follow up in Haiti, he has returned to completely normal function.

Pathological findings

Pathologic examination demonstrated a cyst wall lined by columnar and pseudostratified epithelium with cilia and occasional goblet cells (Fig. 3). These features were consistent with a type A neuroenteric cyst.[6]

Discussion

Neurenteric cysts were first described by Kubie and Fulton in 1928 as “teratomatous cysts”, [7] and later named neurenteric cysts in 1954 by Holcomb and Matson.[8] Neurenteric cysts that come to neurosurgical attention are generally seen in the spine and account for 0.3-1.3% of spinal cord tumors, usually in the cervical or thoracic regions, ventral to the spinal cord.[9, 10] The vast majority, over 90%, are intradural extramedullary lesions.[11] When occurring in the brain, the most common location is in the posterior fossa along the midline, anterior to the brainstem, followed by the cerebello-pontine angle.[2, 12] Unlike spinal neurenteric cysts, intracranial cysts are rarely associated with underlying bony abnormalities such as dysraphism.[13] Intracranial neurenteric cysts are less common in the pediatric population,[3] with the craniocervical junction being a particularly rare location.[4] Dorsally located craniocervical neurenteric cysts are exceedingly rare.[1, 4, 13-20] Previously reported cases with dorsally located posterior fossa or craniocervical junction neurenteric cysts are summarized in Table 1, with only two pediatric cases (<18 years of age at presentation). The current case represents the third reported pediatric case in this location in the literature. The differential diagnosis for dorsally located cysts in this region includes cystic teratoma, arachnoid cyst, or ependymal cyst,[19, 21] all of which are more common.

The average age of diagnosis of intracranial neurenteric cysts is 34 years, despite being a congenital lesion. In contrast, spinal neurenteric cysts tend to present at a younger age (0-10 years).[2] Pediatric cases have also been reported, with a male predilection of 2:1,[22] and a mean age of 6.4 years at presentation.[3] The most common clinical presentation in the pediatric population is spinal cord compression with symptoms of neck pain, myelopathy, and radiculopathy.[3] The spontaneous resolution seen in our case has been described in previous reports, although is not a common feature of this lesion.[10, 17, 23]

Given the unique circumstances due to socioeconomic and access to care barriers, our patient represents an interesting case to demonstrate the prolonged course of waxing and waning of disease with return to normal neurologic state, which would not have been observed in a higher resource setting.

It has been suggested that fluctuations in the cyst size and/or rate of fluid secretion by the columnar epithelium could contribute to the fluctuating degree of spinal cord compression and the waxing and waning clinical picture.[23] Further evidence for the dynamic nature of these cysts is the rare occurrence of recurrent aseptic (Mollaret) meningitis,[24] as can also occur with epidermoid cysts, attributed to the spontaneous rupture of the cyst and leakage of its content into the subarachnoid space.[19, 25]

On MRI, neurenteric cysts are usually isointense on T1-weighted images and hyperintense on T2-weighted images on MRI. However, variations in the protein content of the fluid within the cysts can influence the appearance, especially on T1-weighted images causing them to appear

hyperintense to CSF.[12, 21] Contrast enhancement is generally not seen, and there is usually no solid component.

On pathologic examination, due to the embryologic origin reminiscent of gastrointestinal and respiratory tissue, the neurenteric cyst demonstrates columnar or cuboidal epithelium with or without cilia and intracellular mucous globules.[12, 26] In 1976, Wilson and Odom created a classification of spinal neurenteric cysts based on histopathologic findings with three types: type A is most common, with a single layer of pseudostratified columnar or cuboidal cells; type B includes complex invaginations with glandular organization; and type C which has ependymal or glial tissue.[6]

The embryogenesis of neurenteric cysts is not completely understood, and a number of hypotheses have been proposed to explain the underlying mechanism. These include: split notochord syndrome, persistence of the neurenteric canal, or an aberrant vascular supply to the developing neural tissue.[1, 2, 15] The hypothesis of the persistent neurenteric canal would not completely explain the dorsal location of the cyst in our case and those reviewed in Table 1.[14]

Complete surgical resection remains the gold standard of treatment for neurenteric cysts and is associated with the most favorable outcomes.[2] However, cysts are usually located ventral to the brainstem or the spinal cord, which can hinder complete resection, and therefore recurrence is possible. As a result, postoperative follow-up neuroimaging is necessary to assess the extent of resection and detect any recurrence.[10, 13] Fortunately, the unusual dorsal location of the craniocervical neurenteric cyst in our patient facilitated uncomplicated complete resection through a posterior suboccipital approach. In addition, cyst walls can be adherent to surrounding neurovascular structures, making complete resection potentially hazardous.[4, 14] The literature consensus is that cyst aspiration alone is an ineffective management scheme.[3] It is unlikely to provide histopathological diagnosis, as the cyst fluid is not easily distinguished from other cysts, and the cyst typically recurs since the underlying columnar cellular structure remains preserved.[3] Cyst recurrence has been reported, and is more frequently associated with subtotal resection. As a result, postoperative follow-up is necessary to assess the extent of resection and detect any recurrence.[10, 13]

Conclusion

Neurenteric cysts are rare benign lesions comprised of heterotopic endodermal tissue derived from the gastrointestinal and respiratory tissue. Patients typically present in the second and third decades, but can also present in the pediatric age group. These lesions are commonly located ventral to the spinal cord, but can rarely occur dorsally. Complete resection is curative, and an excellent outcome can be expected after surgery. Our case represents a rare example of a dorsal craniocervical neurenteric cyst with an atypical year-long clinical course due to socioeconomic and access to healthcare barriers, with complete neurologic recovery from severe disability despite persistence of the cyst on MRI.

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Figure Legends

Figure 1. Sagittal T1 (left) and T2 (right) weighted magnetic resonance images showing a dorsally located cystic lesion extending from the level of the medulla oblongata to C3, that is hypointense in T1-weighted image and hyperintense in T2-weighted image.

Figure 2. Postoperative sagittal T1 (left) and T2 (right) weighted magnetic resonance images showing complete resection of the neurenteric cyst via a suboccipital craniectomy and C1 laminectomy.

Figure 3. Hematoxylin-eosin slides showing cyst wall lined by columnar and pseudostratified epithelium (left) and cells with cilia and occasional goblet cells present (right). Original magnification x100 (left), and x400 (right).

Table 1. Review of published dorsally located neurenteric cysts in the posterior fossa or craniocervical junction.

*Discovery reported as incidental, during workup for neck pain.

| Serial No. | Authors | Age (y), Sex | Cyst location | Clinical presentation |
|---------------------------|------------------------|--------------|---------------------------------------------|-----------------------------------------------------------------------|
| Adult patients | | | | |
| 1 | Bolcha et al., 2012 | 27, F | Craniocervical junction to C1 | Dyspnea, dysphagia, nausea, vertigo, paresthesia of lower extremities |
| 2 | Clare et al., 2006 | 57, F | Craniocervical junction | Neck pain |
| 3 | Filho et al., 2001 | 30, F | Craniocervical junction | Ataxia, paresthesia of right hand |
| 4 | Gu et al., 2015 | 39, F | Posterior fossa and craniocervical junction | Intermittent headaches & vertigo, and ataxia |
| 5 | King et al., 2009 | 48, F | Posterior fossa | Neck pain* |
| 6 | Kulkarni et al., 2000 | 40, M | Posterior fossa | Intermittent headaches, progressive ataxia |
| 7 | Macdonald et al., 1991 | 52, M | C1 to C3 | Neck pain, paresthesia of left hand |
| 8 | Weiss et al., 1996 | 32, M | Craniocervical junction to C1 | Meningismus, fever, headache, nausea, vomiting, photophobia |
| Pediatric patients | | | | |
| 9 | Wagner et al., 1998 | 4, M | Medulla oblongata to C2 | Neck stiffness, headache |
| 10 | Zahos et al., 1996 | 17, M | C1 to C2 | Neck pain |
| 11 | presented case | 5, M | Medulla oblongata to C3 | Intermittent episodes of neck pain and transient weakness |





