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Case report

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.

1. Introduction

Neurenteric cysts are uncommon congenital lesions that 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.

2. Case report

2.1. 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. MRI revealed a dorsal cervicomedullary lesion extending to C3 with 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 neck pain. Neuroimaging was repeated and unchanged.

2.2. Neuroimaging

MRI of the cervical spine demonstrated a 2.1 × 2.5 × 4.7 cm, well circumscribed, dorsal intradural, extramedullary cystic lesion extending from the level of the upper medulla to C3 with

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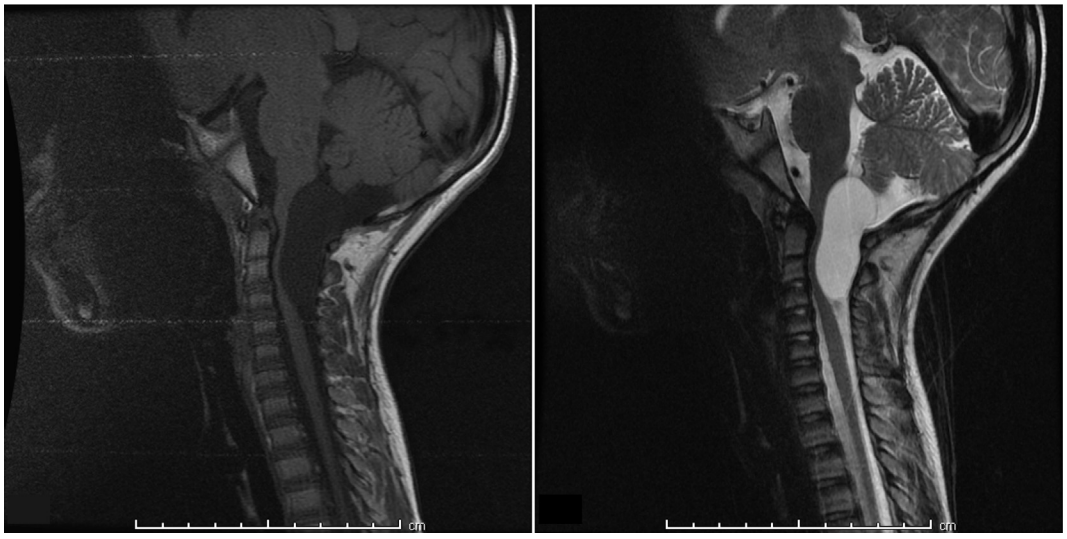

Fig. 1. Sagittal T1 (left) and T2 (right) weighted magnetic resonance images showing a dorsally located cystic lesion extending from the level of the medulla oblongata to C3, that is hypointense in T1-weighted image and hyperintense in T2-weighted image.

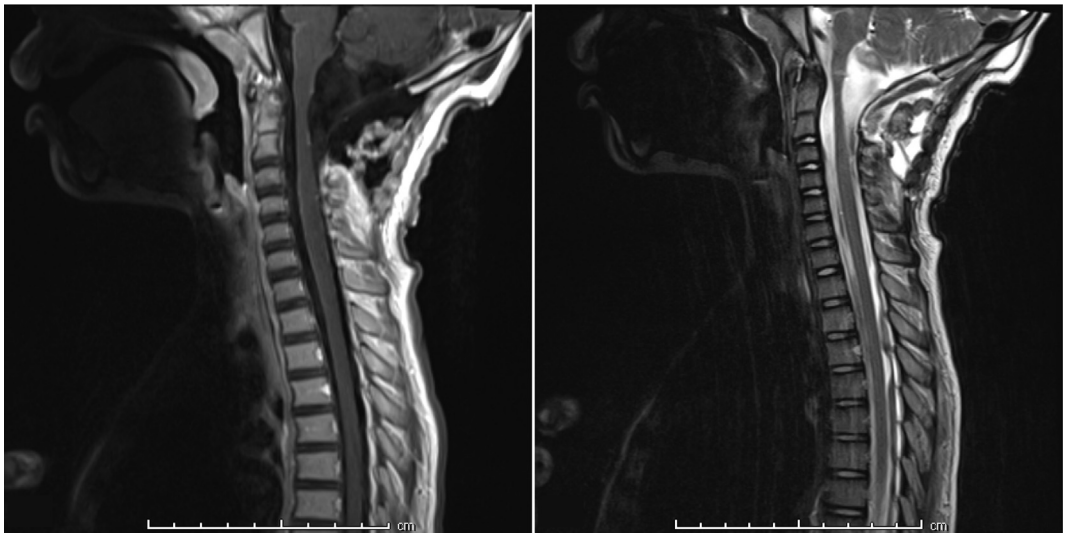

Fig. 2. Postoperative sagittal T1 (left) and T2 (right) weighted magnetic resonance images showing complete resection of the neurenteric cyst via a suboccipital craniectomy and C1 laminectomy.

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). There was no cord edema or associated syringomyelia. There was also expansion of the posterior elements at C1 and C2 to accommodate the cyst, indicating the long-standing nature of the cyst.

2.3. 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.

2.4. Postoperative course

The patient had an uneventful postoperative hospitalization and was discharged home on the 4th postoperative day with no neurologic deficit. At 3 month follow up in Haiti, he has returned to completely normal function.

2.5. 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].

3. Discussion

Neurenteric cysts were first described by Kubie and Fulton in 1928 as "teratomatous cysts", [7] and later named neurenteric

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