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Case Report

Huge Ventral Cervicomedullary Neurenteric Cyst: A Rare Entity with Good Surgical Outcome and Appraisal

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Abstract

Neurenteric cysts are rare congenital lesions of benign nature that can be encountered at any level of the neuraxis, starting from the cranium down to coccyx. Rewarding outcome can be achieved with early diagnosis and complete removal of these benign lesions. Here, we report a case of a huge neurenteric cyst in an 11-year-old boy at the ventral craniocervical junction, a rarely reported entity with literature review. In this article, we focus on the clinical presentation, pathogenesis, radiological findings, surgery, and surgical outcome of this benign lesion, as we succeeded to have gratifying result following surgery in our instance.

Keywords: *Cervicomedullary, magnetic resonance imaging, neurenteric cysts*

Introduction

Neurenteric cysts (NC) are rare congenital lesions arising from the misplaced lining epithelium of respiratory or gastrointestinal tract, commonly found in the posterior mediastinum and often associated with vertebral dysplasia.[1-3] Cervicothoracic junction is the most common location in the craniospinal axis, while intracranial and craniocervical locations are rarer, where they tend to grow ventrally.[4-6] Here, we present a case of an 11-year-old boy having a huge neurenteric cyst ventral to the brainstem, extending from pontomedullary junction down to C2. Appraisal of literature is also deliberated.

Case Report

An 11-year-old boy presented with gradually progressive neck pain and quadriparesis for the past 1 year. Although conscious and oriented, on examination, he was found to have severe neck ache and quadriparesis of Medical Research Council (MRC) Grade 3, rendering him unable to stand without support. His cranial nerves and autonomic functions were intact.

Magnetic resonance imaging (MRI) of cervical spine including craniocervical junction revealed a well-circumscribed, ventrally placed intradural, extramedullary cystic lesion extending from the pontomedullary junction down to C2 measuring 4 cm vertically and 2.5 cm anteroposteriorly. There was a significant mass effect on the cervicomedullary junction and upper cervical spinal cord. The lesion was iso- to hypointense on T1-weighted image (T1WI), hyperintense on T2-weighted image (T2WI) [Figure 1a and b] with a peripheral hypointense rim, displacing the basilar artery posteriorly. There was no appreciable enhancement on postgadolinium sequences or appreciable cord edema and/or syringomyelia.

Image wise, the most likely differential diagnosis of NCs include arachnoid cyst, epidermoid cyst, and dermoid cyst.

With a "hockey stick" incision, a suboccipital craniotomy with opening of the foramen magnum, removal of the C1 posterior arch, and mobilization of the vertebral artery were done to accomplish a far lateral approach. After opening the dura in a curvilinear fashion, a well-circumscribed, encapsulated cyst ventral to the ponto-medullo-cervical region was encountered. After aspiration of dirty white fluid, the extra-axial cyst with a good dissecting plane between the capsule and the surrounding structures, especially the medulla and the cervical cord, was excised en mass.

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The postoperative period was uneventful. The patient started to show signs of improvement from the 3rd postoperative day (POD) by walking with support. His neck pain subsided, and on 7th POD, he was able to stand on one foot. At discharge on 10th POD, he was neurologically intact with MRC Grade 5 power in all four limbs. His postoperative MRI revealed a complete removal of the cyst [Figure 2a and b].

At 2½ years after surgery, the patient was doing well. He was neurologically intact and was attending his school.

Discussion

In 1928, Kubie and Fulton first described neurenteric cyst as ‘‘teratomatous cysts.”[7] Later on, this rare entity was named as neurenteric cyst by Holcomb and Matson in 1954.[8] These cysts are actually considered hamartomas in many previously reported literatures, which is formed as a result of the failure of the separation between the primitive gastrointestinal tract to that of neural crest cells. Due to the pattern of embryogenesis, these lesions are rare in the central nervous system, accounting for 0.7%–1.3% of all spinal cord tumors – usually in the cervical or thoracic regions and ventral to the spinal cord.[9-11] Among them, about 90% are intradural extramedullary in location.[12] Considering the intracranial neurenteric cyst, the most common location is in the ventral brain stem, followed by the cerebellopontine angle.[13] While the spinal NCs are frequently associated with underlying bone abnormalities, intracranial NCs possess a rare association, which is consistent with our reported case.[14]

Male-female ratio of NC is approximately 2:1, and they generally present in the second and third decades of life. Nonetheless, in the pediatric population, the mean age of presentation is 6.4 years. Among them, NCs in the craniocervical junction are a rare occurrence, while the dorsally located craniocervical NCs are extremely rare.[15-19] Although not as rare as the dorsal craniocervical NCs, our male patient of the ventral craniocervical NC presented at an age more than that is average for presentation in the pediatric group.

The majority of the patients present with progressive worsening of the neck or back pain, depending on the location of the cysts. The patient may present with features of myelopathy and/or radiculopathy which depends on the extent and level of compression. In case of ventral brain stem location, there may be additional features of multiple cranial nerve palsies. The clinical course of these cases is usually gradually progressive or may have intermittent relapsing and remitting course because of periodic leakage of fluid content secondary to osmotic and hemodynamic factors or changes in the rate of production and reabsorption by the columnar epithelium the cyst wall or in cases of hemorrhagic events.[20-23] This patient of ours presented with progressive quadriparesis without any remission or relapse, indicating that the cyst had no leakage or intracystic hemorrhage during the course of its growth, and he improved dramatically following surgery. The relapsing and remitting nature of the disease is frequently misdiagnosed as central nervous system demyelinating diseases.[24] Sometimes, traumatic event may initiate or exacerbate such clinical pictures.[25,26] In addition, pediatric patients may also present with signs and symptoms of aseptic meningitis, pyogenic meningitis, pyrexia, bowel bladder disturbance, and paraparesis or paraplegia.[20,27-28]

As in our case, most NCs are usually isointense on T1-weighted sequence and homogenously hyperintense on T2-weighted sequence on MRI. However, variations in the protein content of the fluid influence the appearance on T2WI, causing them to appear hyperintense to cerebrospinal fluid (CSF).[29-31] The cyst was hyperintense than the CSF intensity in T1WI in our case also and that was found to match with the cyst content peroperatively. Contrast enhancement is mostly absent, and there is also no enhancing solid component usually.[31]

On gross appearance, NCs usually appear smooth, regular, contain straw-colored fluid surrounded by a thickened membrane. Peroperatively, we found the cyst to be a well-capsulated one that was well separable from the surrounding structures with a good cleavage plane.

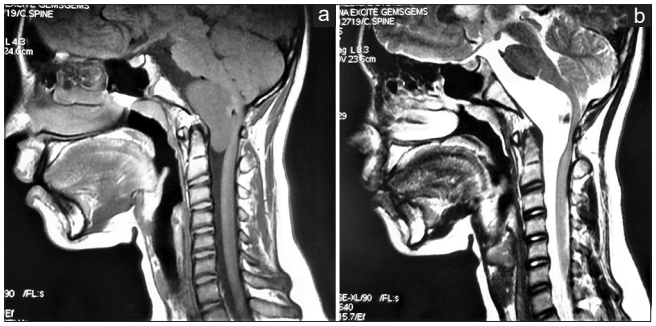

Figure 1: Preoperative sagittal magnetic resonance Imaging showing the ventrally placed intradural, extramedullary cystic lesion extending from the pontomedullary junction down to C2 with significant mass effect on the cervicomedullary junction and upper cervical spinal cord. The lesion was iso- to hypointense in T1-weighted image (a) and hyperintense in T2-weighted image (b)

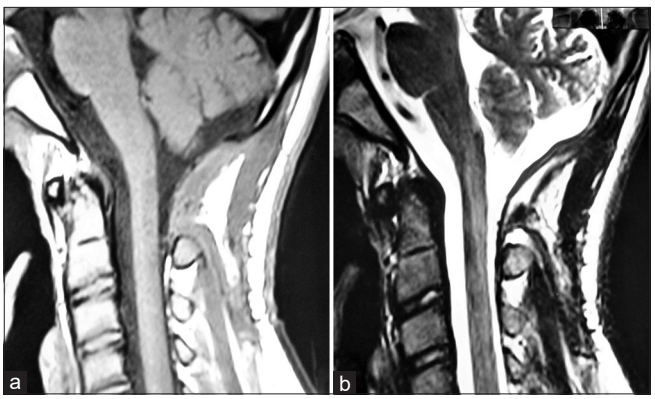

Figure 2: Follow-up magnetic resonance images at 2½ years after showing complete resolution and absence of recurrence in T1-weighted image (a) and T2-weighted image (b)

Moreover, the cyst could easily be excised en mass after aspiration of the dirty white fluid inside. Because of differences in the fluid content, the cyst fluid may have diverse appearances such as “milky,” “blackish,” “CSF-like,” or "clear jelly-like". These varied appearances may result from different degrees of protein contents or from intracystic hemorrhage.[32-34]

On histopathological examination, the neurenteric cyst demonstrates columnar or cuboidal epithelium with or without cilia and intracellular mucous globules, simulating the lining epithelium of respiratory or gastrointestinal tract.[29,35] In 1976, based on histopathology, Wilson and Odom classified NCs into three types: Type A: composed of a single layer of pseudostratified columnar or cuboidal epithelium; Type B: composed of complex invaginations with glandular organization; and Type C: consisting of ependymal or glial tissue.[36] Histopathology of our particular case fell under the category of Type A having cyst wall lined by columnar and pseudostratified ciliated epithelium with the presence of occasional goblet cells [Figure 3].

Complete surgical resection remains the gold standard of treatment for NC, which almost always gives a favorable outcome. In addition to the location of the cyst, the prognosis depends on the size of the cyst as well. As partial resection poses the risk of cyst recurrence, total surgical excision should be the norm, whenever feasible.[27] Depending on location, three basic surgical approaches: posterior, anterior, and lateral can be chosen for cyst excision. In spite of the ventral location, favorable results can be achieved after partial or complete resection of cyst wall through a posterior approach, as reported in different literature.[17,32,37] Now, 2½ years after surgery, our patient is leading a normal healthy life without any recurrence seen in his follow-up MR images [Figure 2a and b]. Although the location was a bit difficult, because of relatively smaller size, good cleavage plane, and following a total excision of the cyst through a far lateral approach, the boy is expected to have a recurrence-free vigorous life. Cysts that are located ventral to the brain stem surgery may be challenging and may result in incomplete resection, which often results in recurrence. Partial resection remains the most important risk factors for the recurrence in NCs, and recurrence rate ranges from 0% to 37% in the literature.[38,39] Because of complete resection, we believe that the boy will not have any recurrence in the future as evident in his postoperative follow-up MR image.

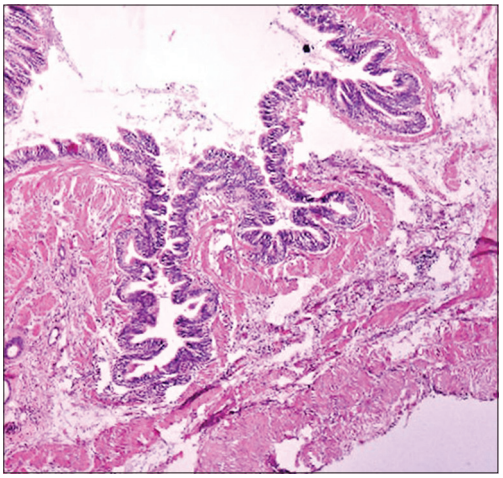

Figure 3: Photomicrograph showing cyst wall lined by columnar and pseudostratified ciliated epithelium with the presence of occasional goblet cells

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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