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Surgical Neurology International

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SNI: Pediatric Neurosurgery

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Case Report

A ventral brainstem neurenteric cyst – A case report and review of the pre-brainstem location

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Received: 16 February 2024
Accepted: 20 May 2024
Published: 07 June 2024

DOI
10.25259/SNI_111_2024

Quick Response Code:

ABSTRACT

Background: Neurenteric cysts are uncommon, benign endoderm-derived lesions that result from aberrant embryologic development of the notochord. They are typically located in the intradural extramedullary spinal cord and rarely located intracranially. Contrary to spinal-located cysts, intracranial cysts are rarer in the pediatric population. Clinically, they may present with symptoms of mass effect, or they can be incidentally discovered.

Case Description: A 10-year-old healthy female child presented with recurrent headaches. The physical and neurological examination was unremarkable. Brain magnetic resonance imaging (MRI) showed a well-demarcated lesion anterior to the pontomedullary junction with striking T1 and T2/T2 fluid-attenuated inversion recovery high-signal intensity and a small rounded nodule within of low signal on T1, T2, and T2*. On initial conservative strategy with serial brain MRI, there was a progressive enlargement of the lesion with significant mass effect on the brainstem. The patient underwent a right retrosigmoid craniotomy, and the cyst wall was fenestrated and drained. Part of the cyst wall and the solid nodule were adherent to the brainstem and basilar artery and were not removed. The histologic findings were consistent with the diagnosis of a benign endodermal cyst. The postoperative period was uneventful.

Conclusion: We report a successful surgical treatment of this rare congenital cyst located in the ventral brainstem. We present pre-and post-operative imaging findings, intraoperative microscopic images of the procedure, and a brief review of relevant clinical literature on the topic.

Keywords: Brainstem, Endodermal cyst, Intracranial neurenteric cyst, Retrosigmoid craniotomy, Ventral

INTRODUCTION

Neurenteric cysts (NECs) are uncommon, benign lesions and are regarded as ectopic endodermal cysts lined by respiratory or gastrointestinal mucin-secreting epithelium. They are thought to arise during the third embryonic week after abnormal or late closure of the neurenteric canal which communicates endoderm and ectoderm through the notochord.[6,11]

They are typically located in the intradural extramedullary ventral spinal cord and rarely located intracranially. Intracranially, they are typically found in the pre-pontine cistern, cerebellopontine cistern, or in the 4th ventricle.[5,13] Contrary to spinal cysts, which are more commonly identified in pediatric patients, intracranial cysts tend to present later and are more often reported in

young adults.[5] When intracranial, they are either discovered incidentally or following symptoms that vary from headaches to symptoms resulting from compression of the brainstem (BS) or cranial nerves. The radiological diagnosis is not straightforward due to variable protein content within the cyst, resulting in variability in T1 and T2 magnetic resonance imaging (MRI) signal and also due to alternative similar lesions.[10]

The best surgical management remains controversial due to the rarity of the condition and the limited follow-up data. Complete resection of the NEC, including the cyst wall, is generally recommended.[4,9,13] However, the intricacies of this location with a potentially hazardous attempt at removal demand considering a different approach.

We present a case of a 10-year-old child with an NEC located ventrally to the BS, which showed progressive growth in follow-up exams. We present the pre-and post-operative imaging findings and a brief review of the relevant clinical literature on the topic.

CASE DESCRIPTION

History and imaging

A 10-year-old healthy female child presented with recurrent occipital headaches over 1 year**.** The physical and neurological examinations were unremarkable. Head computed tomography imaging showed a small, rounded, hyperdense lesion located anterior to the pontomedullary junction. On MRI, the lesion seemed extra-axial and was mostly hyperintense on T1 and T2/T2 fluid-attenuated inversion recovery, with facilitated diffusion, no chemical-shift artifact (not a fatty lesion), and no gadolinium enhancement, with a small nodule within of low signal on T1, T2, and T2*, suggestive of calcification. The lesion measured about 10 × 9 × 8 mm in size (anterior-posterior × transversal × craniocaudal) and 280 mm^3 of volume and had no surrounding edema, with a slight mass effect on the BS [Figure 1]. Due to the relatively innocent clinical manifestations and dimensions of the lesion, a "watch and wait" strategy was initially employed.

On an initial conservative strategy with serial follow-up brain MRIs, there was a progressive growth of the lesion, which became more lobulated and extended to the pre-bulbar and pre-pontine cisterns, measuring 19 × 28 × 23 mm and 5400 mm^3 of volume, with significant mass effect. The prepontine vasculature was further anteriorly displaced but not encased. This led to the decision of neurosurgical treatment [Figure 2].

Operative details

The patient was positioned in a lateral position, and we performed a right retro mastoid craniotomy with an extension to the foramen magnum. On an operating microscope and neuronavigation-assisted, the cyst wall was exposed, widely

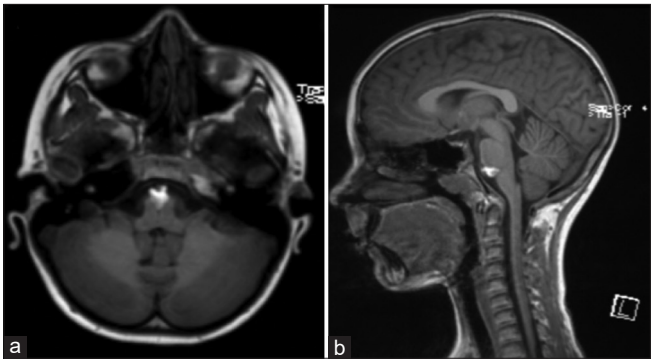

Figure 1: Brain magnetic resonance imaging at diagnosis. (a) Axial T1 and (b) sagittal T1 show a spontaneously hyperintense mass anterior to the pontomedullary junction.

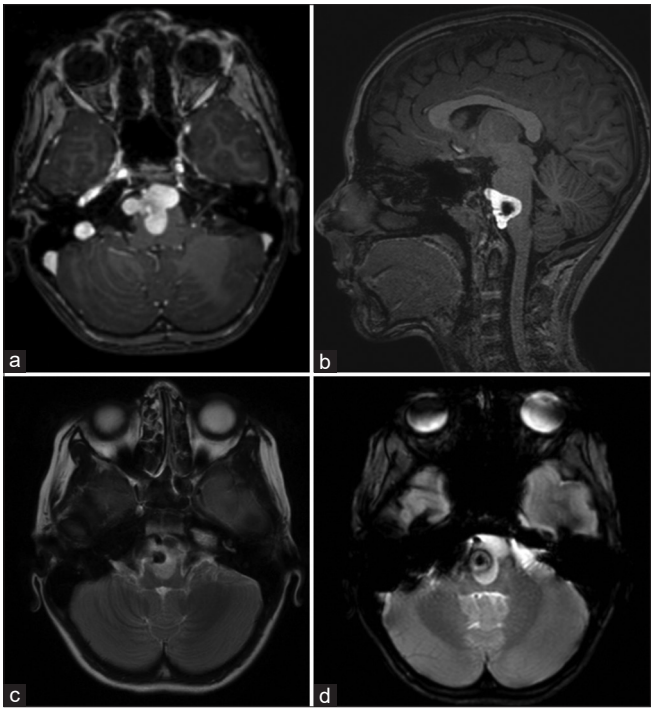

Figure 2: Brain magnetic resonance imaging on a 2-year follow-up, showing an increase in lesion volume and mass effect on the brainstem. A small nodule within of low signal on T1, T2, and T2* is apparent. An anteriorly displaced but not encased basilar artery is also visible (a) sagittal T1, (b) sagittal T1 with contrast, (c) axial T2, and (d) axial T2.

fenestrated, and its fluid was drained, revealing a thick yellow liquid content. After aspiration and profuse irrigation of the cyst, part of the cyst wall was removed. The part of the cyst wall adherent to the BS and basilar artery was not removed [Figure 3].

Histopathology and postoperative details

The histologic findings were of a pseudo-stratified epithelium with no anaplasia, consistent with the diagnosis of a benign

endodermal cyst. In the postoperative period, the patient experienced no neurological alterations, and postoperative MRI showed complete drainage of the cyst and the persistence of a small T2 hypointense nodule. At 1 year follow-up, the patient was well, active, and with no symptoms, and imaging showed no evidence of recurrence of the lesion [Figure 4].

DISCUSSION

Intracranial NEC accounts for 0.01% of central nervous system tumors.[5] They tend to be located anterior to the BS and commonly extend into the cisterns to produce a mass effect in the ventral BS. The cranial presentations are rarer in the pediatric population.[5,9] We conducted a search in the National Library of Medicine with the keywords "neurenteric cyst" and "brainstem." Of the 1689 results, we found 40 publications, including posterior fossa NEC. Reports on CPA or NEC posterior to the BS were excluded from the study. Only seven reports described treated BS or pre-BS NEC in the pediatric population (0–18 years) [Table 1].[1,9,12,14]

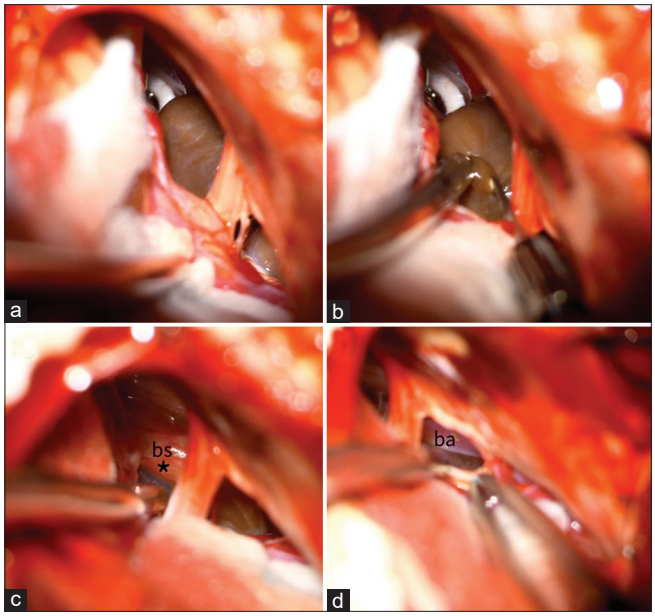

Figure 3: (a) Exposure of the lesion's capsule through the cerebellopontine angle between the 5th and the 7th nerve and its inferior part of the cyst viewed between the 7th and the 9th nerves, (b) the capsule was opened, and a dense yellow fluid was aspirated, and (c and d) final view after fluid drainage and partial removal of the capsule, part of the capsule (*) left adherent overlying the brainstem (bs). The basilar artery (ba) is also viewed.

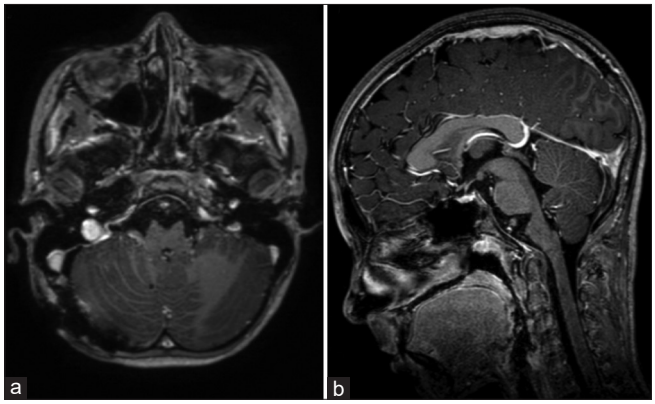

Figure 4: Postoperative brain magnetic resonance imaging T1 sequence with gadolinium (a) axial view and (b) sagittal view showing no evident residual cyst and resolution of the mass effect on the brainstem.

In the presented case, despite the significant growth of the lesion, the patient had no neurological signs, which we relate to the slow growth of the cyst to the prepontine cistern. This contrasts with some cases that reported quicker growth.[12] This case was remarkable for the mass effect and the absence of symptoms of BS or cranial nerve compression. This posed an additional difficulty in the surgical decision due to the risk associated with an approach to this region in a neurologically intact child.

Despite its rarity, the imaging features of an intracranial NEC can be suggestive of the diagnosis. NEC can present with a variety of different signal intensities on T1 and T2 related to differences in protein levels, rendering the diagnosis challenging.[7] Differential diagnosis should include other cystic intracranial lesions, such as dermoid and epidermoid cysts, and even tumors (such as pilocytic astrocytoma). The location adjacent to the ventral BS, at a pontomedullary level, along with the signal evolution on MRI, can give a precious clue to this diagnosis. In particular, the non-gadolinium enhancement of the facilitated diffusion, along with a T2/T2* hypointense nodule inside the lesion, are evocative of the diagnosis.

The existence of a nodule has been attributed to calcification or previous hemorrhage, but in a recent report of removal of such an associated nodule, the histologic analysis showed a xanthogranulomatous reaction with melanin. It has been proposed that this nodule/hemorrhage favors inflammation, which might explain the transformation from a silent cyst to an active cyst with headache and cyst expansion.[15]

Complete surgical resection is the gold standard for the treatment of NEC and is associated with the most favorable outcomes.[4,13] However, from our review, it is shown that subtotal resection is more frequent than total resection in this location. In our case, the cyst wall was opened, the cyst drained, and copiously irrigated. Part of the cyst wall and the solid nodule were not removed due to adherence to both the BS and vascular structures.

Despite a 1-year follow-up MRI showing no signs of recurrence, a long follow-up is recommended since an age under 30 years old is a risk factor for recurrence.[4] An up to 37% recurrence rate has been reported during a span of weeks to 14 years for NEC in general and specifically for pre-pontine cysts, an 11-year recurrence has been reported.

Table 1: Case reports on brainstem neurenteric cyst (NEC).

Case	Reference	Number of cases	Age	Clinical presentation	Location	Intracystic nodule	MRI T1 signal	Extent of resection	Duration of recurrence
1	Oliveira *et al.* 2005[9]	2	5, 11 yr	Diplopia Incidental	Ventral to brainstem (Clivus)	NA	NA	Total	NR at 1 and 4 yr of FU
2	Ko *et al.* 2008[8]	1	4	Intermittent headache and vomiting	Ventral brainstem	Yes	Hypo	Total	NA
3	Birinyi *et al.*, 2014[3]	1	4	Gait ataxia, Facial Palsy	Ventral brainstem	NA	Hypo	Total	NR at 6 mo of FU
4	Wong *et al.*, 2016[14]	1	16 mo	Incidental at first, however, progressing to right CN VII palsy, diplopia, and hemiparesis	Intra-axial brainstem	Yes	Hypo	Subtotal (cyst drainage and nodule removal)	R at 15 mo, NR at 9 mo FU after 2nd surgery-intracystic H2O2
5	Shimizu, *et al.*, 2019[12]	1	7 yr	H/A, Diplopia, dysarthria CN VI and VII palsy	Intra-axial brainstem (pons)	Non-apparent	Hypo	Subtotal	NR at 5 yr of FU
6	Agresta, *et al.*, 2020[1]	1	16 mo	Vomiting and swallowing difficulty, developmental regression, left CN VII palsy.	Intra-axial brainstem	Non-apparent	Hypo	Subtotal (only cyst drainage)	R at 3 wk, NR at 9 mo FU after 2nd surgery-intracystic IFN-Y
7	Current reported case	1	10 yr	H/A	Pre-brainstem	Yes	Hyper	Subtotal (drainage and partial cyst wall resection)	NR at 1 yr of FU

CN: Cranial nerve, H/A: Headache, H_2O_2: Hydrogen peroxide, FU: Follow-up, IFN-α: Interferon α, mo: Month/s, NA: Non-applicable, NR: Non-recurrence, N/V: Nausea and vomiting, R: Recurrence, wk: Week/s, yr: Year/s, MRI: Magnetic resonance imaging

However, recurrence is more frequent in the first 2 years.[1,2,4]

In recurrent cases, different approaches have been reported. Newly formed adhesions may complicate reoperation with new drainage and further attempt to remove the cyst wall. A case of an intra-axial BS recurrent NEC was treated with intra-cystic Interferon-α and another with intra-cystic H_2O_2, with good outcomes.[1] In other rare cases, the implantation of a shunt was also reported.[4]

CONCLUSION

We report a surgical resection of this rare congenital cyst located ventral to the BS. This case adds to previously reported cases on the management of prepontine NEC in which adherence to critical structures precludes total removal. It also evokes questions on the natural history of the lesion and the causes of growth. More data on this pathology and its natural history are needed to optimize management and recurrence avoidance.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the

writing or editing of the manuscript and no images were manipulated using AI.

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How to cite this article: Lemos SS, Sá MI, Casimiro C, Guerreiro C, Faria C. A ventral brainstem neurenteric cyst – A case report and review of the pre-brainstem location. Surg Neurol Int. 2024;15:195. doi: 10.25259/SNI_111_2024