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Neuroanatomical studies

Surgical anatomy of a neurenteric cyst anterior to the brainstem

Marcelo Budke[a], Davide Luglietto[b,*], Francesco Vitaliano Muzii[b]

[a] Department of Neurosurgery, Hospital Infantil Universitario Niño Jesús, Madrid, Spain
[b] Department of Medicine, Surgery and Neuroscience, Section of Neurosurgery, University of Siena, Siena, Italy

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ABSTRACT

The authors report on a sixteen-month old boy who presented a neurenteric cyst located in the cervicomedullary junction, anterior to the brain stem. Centred on the ventral aspect of the premedullary cistern, the cyst extended from the prepontine space to the cervicomedullary junction, causing important brain stem compression. The lesion was totally removed via a posterior approach with suboccipital craniotomy and laminectomy of C1. After surgery the patient recovered his neurological function and the postoperative MRI showed total resection of the cyst.

1. Introduction

A neurenteric cyst is an uncommon congenital lesion first described by Puusepp in 1934 [1,2]. This type of cyst is generally believed to be the result of the split notochord syndrome [3]. It is derived from displaced endodermal tissue and most commonly encountered in the posterior mediastinum.

They are quite rare in the central nervous system, but are sometimes found in the intradural extramedullary space in the lower cervical and upper thoracic spine [1,4,5]. Intracranial neurenteric cysts are even rarer and most of them are found in the posterior fossa [6–8].

We describe a new case of neurenteric cyst that is anterolateral to the brain stem and briefly discuss the surgical management and features of these rare intracranial cysts.

2. Case report

2.1. History and examination

A sixteen-month old boy was brought to Hospital with poor gait and left arm weakness and torticollis. Seven weeks before he had developed torticollis with left deviation of the head. Two weeks later he lost strength in his left arm. In the two weeks prior to admission, his gait became clearly affected. Neurological examination revealed a mild motor deficit in his left arm and hand (4/5).

2.2. Neuroimaging findings

The CT and MRI revealed a 3 × 1.5 cm cystic lesion extending from to the prepontine space to the cervicomedullary junction on the ventral aspect of the premedullary cistern. The mass of the cyst was compressing and distorting the brain stem (Figs. 1 and 2). The lesion was hypointense on T1-weighted images and hyperintense on the T2-weighted and FLAIR images. No enhancement was observed after Gadolinium administration.

2.3. Operation and postoperative course

Placed in a prone position, the patient underwent a suboccipital craniotomy via a left laterocervical approach with laminectomy of the C1 posterior arch (Fig. 3). After lateral opening of the dura and arachnoid membrane under microscopic magnification, the capsule of the cyst was observed lying between the vertebral artery, the intradural rootlets of C1, and the cervical spinal cord (Fig. 4). The large cyst was completely resected “en bloc” after gentle retraction of the capsule. There was no mucus within the bubble cyst and the content was transparent, mimicking a hydatid cyst. After surgery, the patient made a good recovery and his motor weakness rapidly disappeared within two weeks. The postoperative MRI revealed total resection of the cyst and normalization in the anatomy of the pons and medulla (Figs. 5 and 6). At three years postsurgery the child is developing normally with no neurological or motor deficits or signs of reappearance of the cyst.

2.4. Histopathological examination

The 1.5 cm diameter cyst was sent to pathology. Grossly, the cyst had a thin, semitransparent wall that contained a translucent liquid. The tissue was fixed in 10% neutral formaline and embedded in

* Corresponding author.
E-mail address: davidelugliettomd@gmail.com (D. Luglietto).

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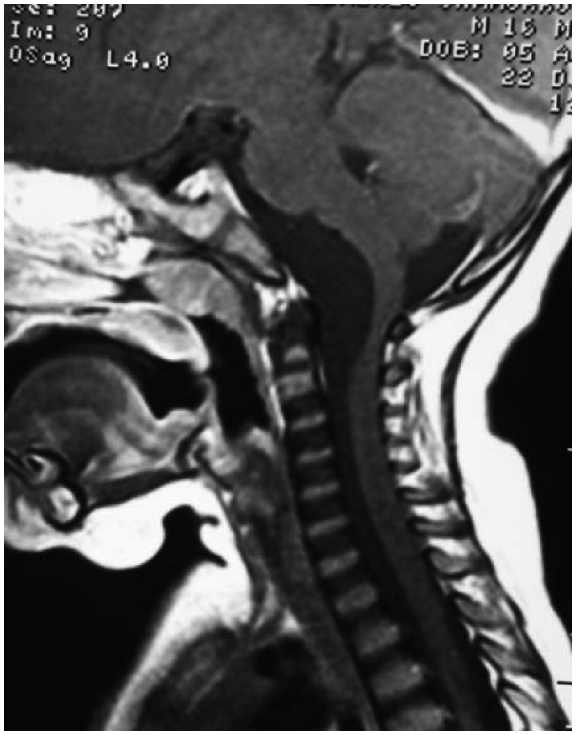

Fig. 1. Sagittal T1-weighted MR image illustrating a hypointense mass anterior to the brain stem.

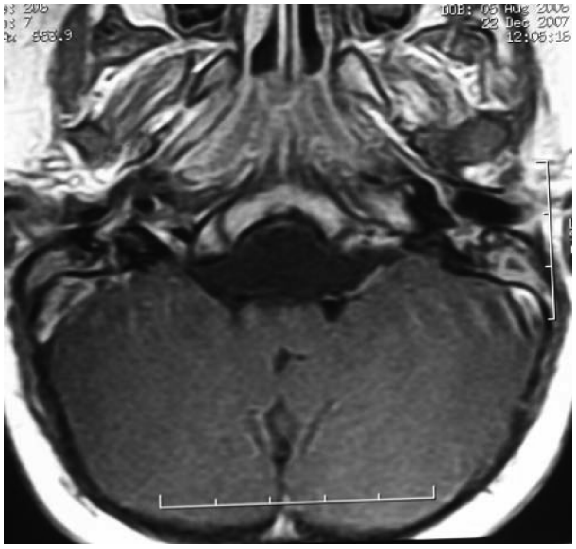

Fig. 2. Axial T1-weighted MR image illustrating a hypointense mass anterior to the brain stem.

paraffin. Three micra-thick sections were stained for hematoxylin-eosin. Histological examination of the cyst wall showed a simple columnar epithelial cyst wall without ciliated cells resting on a thin layer of fibrous connective tissue, consistent with the diagnosis of neuroenteric cyst (Figs. 7 and 8).

3. Discussion

Neurenteric cysts, also known as endodermal, enterogenous, gastrogenic or gastroenterogenous, gastrocytomas, archenteric or foregut cysts, teratomas or intestinomas [9], are usually encountered as a right mediastinal mass on a child being evaluated for respiratory or swallowing symptoms [10]. However, they have occasionally been

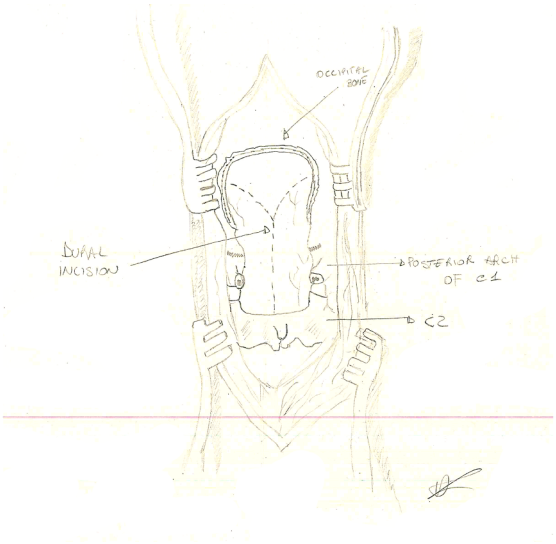

Fig. 3. Sketch of the craniotomy and dural exposure.

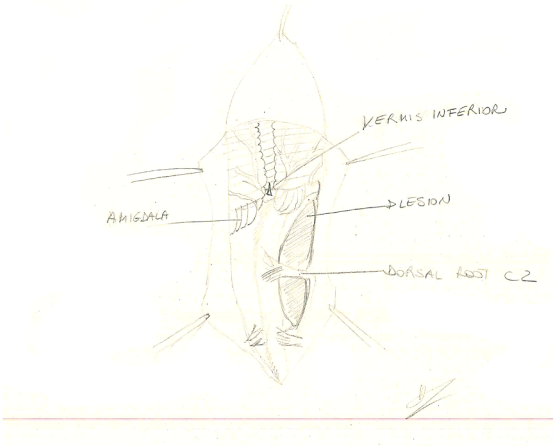

Fig. 4. Sketch of the surgical field after dural opening and exposure of the cyst.

described affecting the central nervous system in regions from the cerebellopontine angle to the coccyx [11–13]. In these circumstances, they are typically found during the second and third decades of life, and located in the spinal canal at the lower cervical and upper thoracic segments, commonly in intradural and extramedullary spaces, anterior or anterolateral to the spinal cord. More than one hundred cases of intraspinal location had been reported between 1926 and 1995 [14].

The intracranial location of neuroenteric cysts is extremely rare, and only a few cases have been published to date. Posterior fossa neurenteric cysts constitute more than 90% of the intracranial form of these cysts, and they occur mostly along the midline (in the fourth ventricle and prepontine cistern, or within the brain stem), although they may also be seen laterally or within the cerebello-pontine angle (CPA) cistern [8,15–18]. In a review by Bejjani et al., the most common locations of posterior fossa neurenteric cysts were anterior to the brainstem (51%), within the fourth ventricle (21%), extending into the cervical region (18%), and in the CPA (17%) [6].

These lesions are frequently incidental neuroimaging findings, but they may occasionally produce symptoms of progressive neurological impairment and chronic increased intracranial pressure [19]. The symptoms of a posterior fossa neurenteric cyst can be due to an inflammatory reaction or a mass effect. Active secretion by goblet cells can result in cystic expansion and compression of neighbouring neurovascular structures [7]. The most frequent complaint is headache,

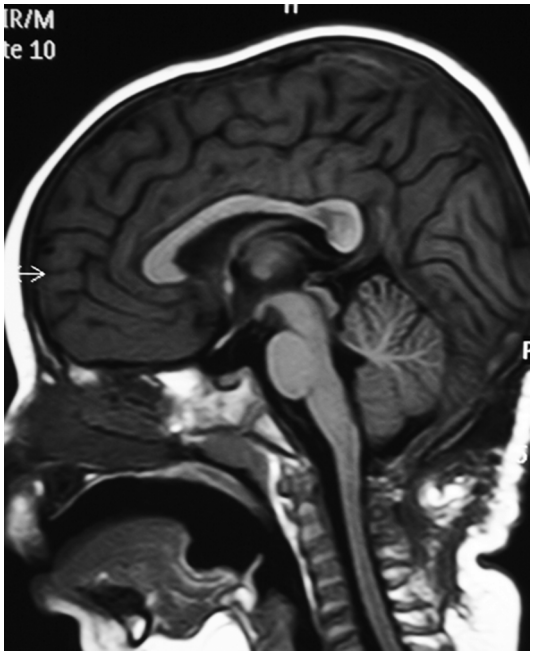

Fig. 5. Sagittal T1-weighted MR image in the early postoperative period revealing total removal of the cyst.

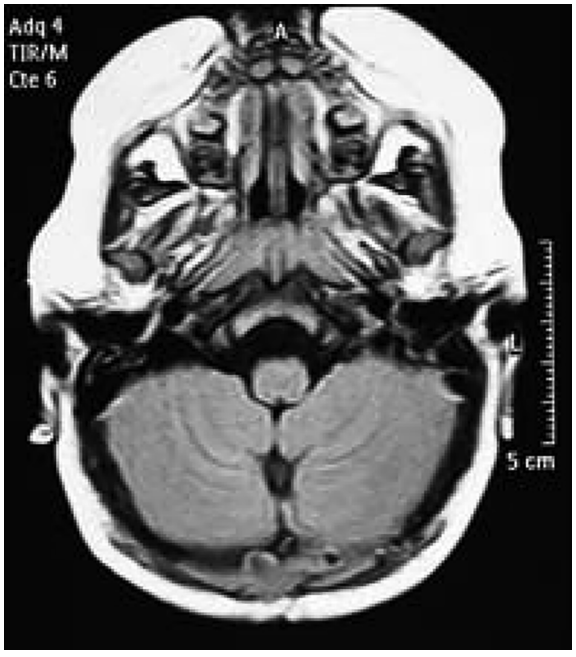

Fig. 6. Axial T1-weighted MR image in the early postoperative period revealing total removal of the cyst.

followed by gait disturbance and motor and sensory disturbances [6]. Involvement of the cranial nerves can result in facial numbness, hemifacial spasm, hearing loss, and swallowing difficulties. The intermittent cyst leakage or spontaneous rupture can result in recurrent aseptic meningitis and/or hydrocephalus secondary to chronic meningitis [20,21]. In the present case, the ventral compression of the brain stem and upper cervical cord were the origin of the child's neurological deficit.

The variability in the imaging characteristics of neurenteric cysts has been attributed to the presence of proteinaceous or other hydrophilic contents [19]. Computerized tomography scanning usually reveals a hypodense lesion, although the lesion occasionally can be

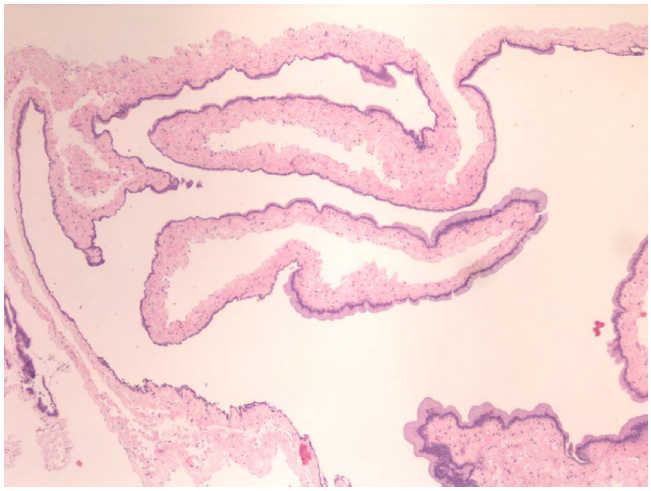

Fig. 7. Cyst wall and lumen which shows columnar epithelium on fibrous tissue (40 × H.E.).

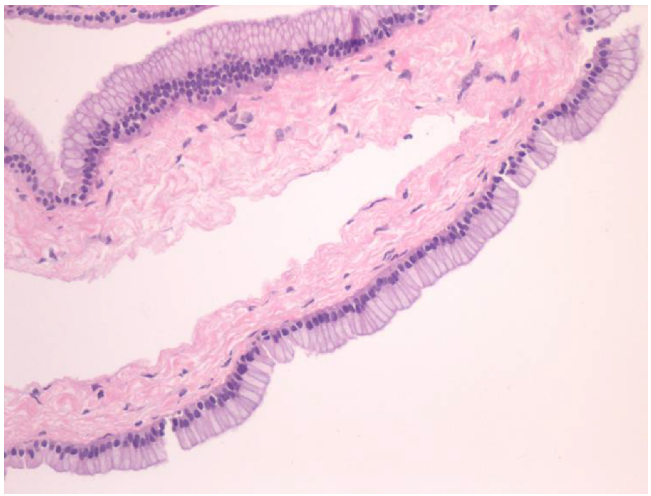

Fig. 8. Benign epithelium columnar resting on a thin layer of fibrous tissue (200 × H.E.).

hyperdense. MRI is the study of choice for of neurenteric cyst diagnosis. These lesions may appear hypointense, isointense, or slightly hyperintense on T1-weighted MRI images and mostly hyperintense on T2-weighted and Flair images. Usually the cyst wall does not enhance after gadolinium injection [20]. In our case the cyst was hypointense on T1-weighted images and hyperintense on both T2-weighted and FLAIR images.

Neurenteric cysts are benign lesions that are believed to originate in embryonic disgenesia [1,3,14,19]. There are several hypotheses about the pathogenic mechanism behind their development. Neurenteric cysts form during early embryogenesis as the result of some disturbance in the interrelations between notochord, neural tube, endoderm, and mesenchyme. Disruption of this process may lead to the inclusion of endodermal tissue and cystic formation, so the embryogenesis of a neurenteric cyst probably dates from the first three weeks of gestation, when the three embryological layers are in close proximity.

Pathologically, these cysts vary in composition and complexity. The simplest cases are lined by a single layer of simple, pseudo- or fully stratified cuboidal or columnar epithelial cells, with or without cilia, lying on a basement membrane and supported by a layer of more or less vascular connective tisuue [9,22]. The cyst may have different compositions. The fluid may be clear, mucoid or xanthochromic [8]. A dark brown colour is indicative of intracystic haemorrhage [23]. In our case,

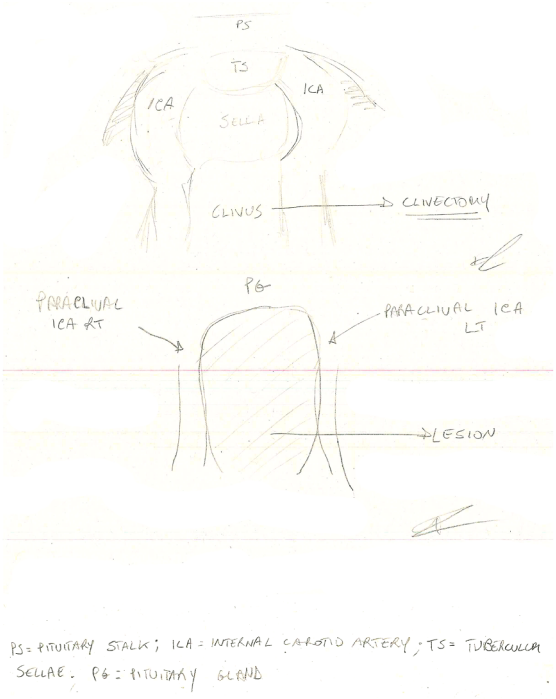

Fig. 9. Sketch of the endoscopic endonasal transclival approach to the lesion.

the cyst had a firm capsule and a clear content.

The treatment of a neurenteric cyst entails the complete excision of both the cyst contents and the cyst [6–8]. However, in some cases, portions of the cyst should not be resected because of their location or adherence to surrounding vital structures. In these cases, subtotal removal is a reasonable alternative to avoid damaging these structures. Aggressive surgical manipulation to detach a firmly adhering cyst may result in severe neurological deficits and should be avoided. In our case, the cyst resection was simple and complete. It was removed from the adjacent neural tissue using a posterior approach with gentle traction on the cyst.

Some authors, like Arai [24], Menezes [25], and Devkota [4], have recommended an anterior surgical approach for the complete surgical resection of ventrally-located cervicomedullary intradural extramedullary neurenteric cysts (Fig. 9), while others have advocated a far-lateral transcondylar approach (Liu and Couldwell, 2005). On the other hand, Park at al. [1] stated that surgery using a posterior approach, such as a laminotomy or laminectomy is sufficient for decompression and total removal of the cyst, whether it is ventrally or dorsally located. The present case supports the latter proposition. In our opinion the best surgical approach for any individual patient is the one that is safest and most effective for that patient.

4. Conclusions

We have reported a very unusual paediatric case of neurenteric cyst at the cervicomedullary junction. This challenging lesion was successfully resected using a suboccipital approach and careful microsurgical dissection. More sophisticated skull base techniques were unnecessary to achieve complete surgical removal.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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