



One and a Half Syndrome Complicating Resection of Fourth Ventricular Neurenteric Cyst

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Abstract

Neurenteric cysts are rare benign lesions of the central nervous system, typically occurring anterior to the brainstem. They are thought to arise from disturbances in early gastrulation. A neuroenteric cyst, appearing wholly within the fourth ventricle without obvious connection to the ventral brainstem, is described. Removal of the cyst caused a one and one half syndrome, which slowly resolved after surgery. Reoperation on recurrent cyst showed a midline raphe in the fourth ventricle, leading the authors to hypothesize that this raphe represented a gliotic band from a split notochord during development, which may have transmitted vibration during cyst removal, leading to brainstem injury and the resultant one and one half syndrome.

Keywords: Fourth ventricular neurenteric cyst; Gastrulation

Introduction

Neurenteric cysts are uncommon lesions of the central nervous system. They consist of epithelial cells and may contain mucosal glands, cilia, and other such features common to the digestive or respiratory tracts. Their diagnosis can be confusing as they can easily be misdiagnosed as arachnoid or epidermoid cysts [1-3]. Due to this confusion, they are known by many names throughout the literature such as endodermal cysts, enterogenous cysts, intestinomas, gastrocytomas, gastroenterogenous cysts, or archenteric cysts [2,4]. According to Gauden et al. [1], only 140 cases of intracranial cysts have been reported since 1952 [1]. Intracranial cysts are most commonly located in the posterior fossa [3,5,6]. The most common posterior fossa locations are anterior to the brainstem in the cerebellopontine angle and within the fourth ventricle [5-7]. They are rarely positioned supratentorially [1,2].

It has been hypothesized that neuroenteric cysts arise from defects in gastrulation. Since neuroenteric cysts represent cells of endodermal origin, it has been postulated that herniation of an endodermal layer during gastrulation through a “split notochord” may allow the adhesion of cells of endodermal origin into developing neural folds of the ectoderm [8-10]. This hypothesis would show how cells could be left behind within the fourth ventricle to form neuroenteric cysts without obvious ventral connections but does not fully explain how a “split notochord” may re-join without the development of a gliotic median raphe along the midline during brain development. This illustrative case report will suggest that the complications arising from the removal of a dorsally located neuroenteric cyst might best be explained by a ventral connection to endodermal tissue along a gliotic midline raphe.

Case Report

A 57-year-old man presented with vertigo when he turned his head to the left for associated nausea and vomiting. The patient's physical exam and neurological exam were normal. An MRI, revealing a cystic lesion dorsal to the brainstem and splitting the cerebellar hemispheres, appeared to follow CSF on all sequences (Figure 1A-D). A workup for possible neurocysticercosis was negative, leading to a clinical and radiographic diagnosis of arachnoid cyst.

The patient underwent sub occipital craniotomy with microsurgical resection of the lesion. After opening the dura, a multiloculated cystic mass was identified located inferior to the cerebellum and extending into the fourth ventricle resembling a mass of grapes. Cystic masses were dissected from the fourth ventricle with minimal difficulty with no bleeding nor breach of the pia of the floor of the fourth ventricle.

After surgery, the patient complained of double vision. An MRI failed to show either diffusion

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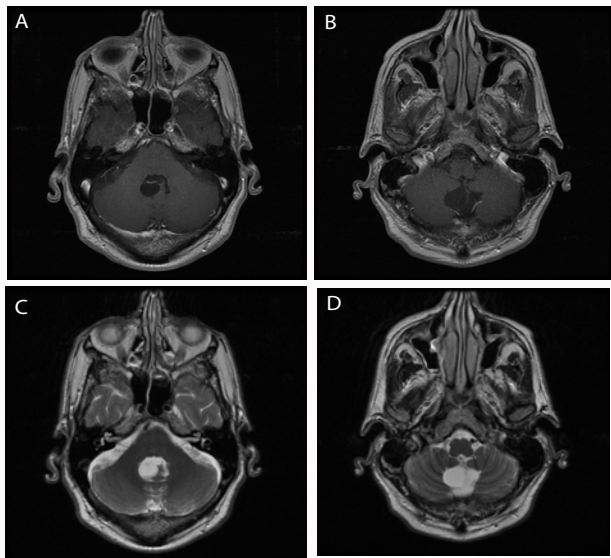


Figure 1: Preoperative axial T1 weighted with gadolinium (A,B) and T2 weighted axial images (C,D) through the lesions. Note the cystic structures between the tonsils and the cerebellum extending into the fourth ventricle.

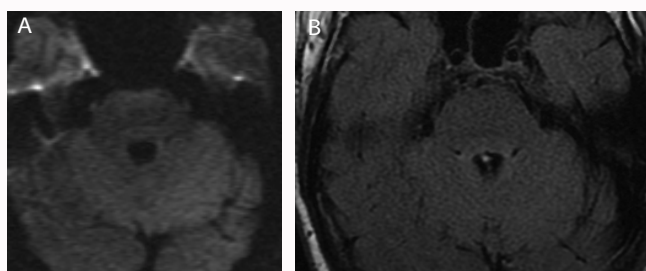


Figure 2: Postoperative diffusion weighted (A) and FLAIR (B) images through the floor of the fourth ventricle at the level of the PPRF. Note that there appears to be no swelling or ischemia noted postoperatively at the level of the expected injury given the clinical findings.

weighted imaging or FLAIR changes suggestive of stroke or other injury to the pons (Figure 2A and B). He was diagnosed with having internuclear ophthalmoplegia and a left sixth nerve palsy. Mild imbalance rapidly occurred when turning his head. The patient was prescribed steroids and given an eye patch to help correct the issue. A multicystic lesion with mucoid material, mucinous epithelium, and basement membranes was identified on pathological examination (Figure 3A and B). Epithelial membrane antigen (EMA) was focally positive, and a diagnosis of neuroectodermal cyst was made.

Although his internuclear ophthalmoplegia (INO) and sixth nerve palsy improved at seven months postoperatively, he still had a subtle right INO and a slight left gaze (sixth nerve) paresis, representing the infrequent diagnosis of one-and-a-half syndrome.

Shortly thereafter, a follow up MRI showed the re-growth of a cyst in the fourth ventricle, and a second operation was performed (Figure 4A-D). The lesion, a purplish-hued cyst, was situated at the floor of the fourth ventricle (Figure 5A). It was separated from the brainstem using micro dissection techniques. A subtle midline raphe and gliosis were noted on the floor of the fourth ventricle between the facial colliculi (Figure 5B). The patient tolerated the second procedure well without obvious worsening of his subtle INO and sixth nerve palsy.

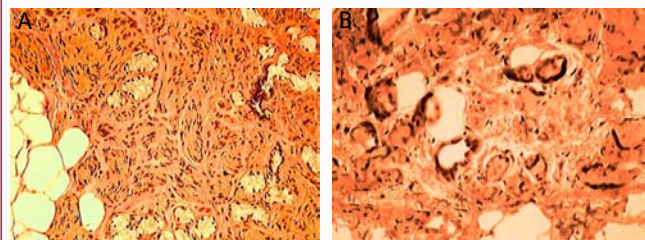


Figure 3: H&E stains (A) and EMA stains (B) of the resected cyst. Note the cuboidal epithelium present in (A) and the brown staining of the EMA along the cuboidal epithelium (B).

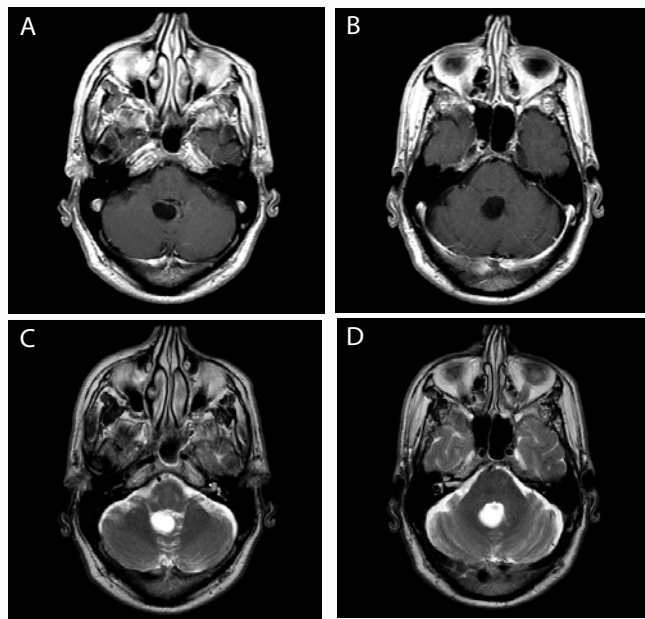


Figure 4: Preoperative axial T1 weighted with gadolinium (A,B) and T2 weighted axial images (C,D) through the recurrent cystic lesion. The cystic structure now appears to be a single cyst higher in the fourth ventricle than the prior cysts.

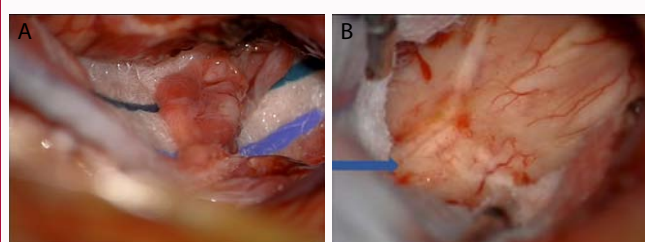


Figure 5: Intraoperative microscopic view of the cyst wall (A) resting on an underlying cottonoid. After removal of the cyst, the floor of the fourth ventricle is visible (B). The arrow points to the midline raphe to which the cyst was attached and is postulated to be the source of the injury to the patient during the first cyst removal procedure.

Discussion

Neurenteric cysts are benign central nervous system lesions. They account for just 0.01% of central nervous system tumors [1]. When these cysts do occur, they are frequently sited ventral to the spinal cord with spinal cases occurring about three times more frequently than intracranial ones [4]. Intracranial neurenteric cysts are generally traced to the posterior fossa, either anterior to the brainstem in the cerebellopontine angle or in the fourth ventricle. [5-7]. There have

also been a small number of cases reported supratentorially [1-3].

Although spinal neurenteric cysts are most common in pediatric patients, intracranial neurenteric cysts happen in all ages with an approximate median age of 34 [1]. While some studies have determined a male predominance, [3,5] other studies have found a female predominance [4]. Therefore, most authors conclude that neurenteric cysts are independent of sex [1,4,7].

Classification

Neurenteric cysts consist of epithelial cells of a presumed endodermal origin. They may or may not be ciliated, and they lie on a basement membrane, a key feature that differentiates neurenteric cysts from the very similar neuroepithelial cysts [11]. Wilkins and Odom developed a method for classifying neurenteric cysts based on histological features [11]. Type A cysts are lined with pseudo stratified cuboidal or columnar epithelial cells that are similar in appearance to respiratory or gastrointestinal cells. Type B cysts may additionally have complex invaginations and have glands for producing mucinous or serous fluid. These cysts may also contain additional cell types that are associated with gastrointestinal or respiratory tissue such as smooth muscle, lymphoid tissue, and striated muscle. Type C cysts, in addition to the features of type A and B cysts, hold glial elements.

Imaging

There is a great deal of variance in the imaging of these cysts, most likely due to the variable protein makeup of the cysts' contents [2,12,13]. Cysts most usually appear as hypo dense on CT scans, but they may also appear as hyper dense [2,4,5]. Contrast enhanced CT scan rarely show enhancement, however, some cases do exhibit enhancement [4,5,7]. On MRI, cysts typically appear as is intense to hyper intense to CSF on T1 weighted MRI [1,4,7,12,14]. They usually seem hyper intense to CSF on T2 weighted images [4,7,12]. Since neurenteric cysts have imaging properties very similar to other kinds of intracranial lesions, the diagnosis of a neurenteric cyst by radio imaging alone is quite challenging.

Histopathology

Immuno histochemical analysis is the best way to definitively distinguish a neurenteric cyst from other entities. Due to their endodermal origin, neurenteric cysts are generally negative for both GFAP and S-100 [2-3,6,7]. They are positive for many endodermal markers such as cytokeratin, EMA, and carcinoembryonic antigen [2-3,6,7] and are often positive for acid Schiff staining confirming the presence of goblet cells [1,3,7].

Case Discussion

This case represents a rare occurrence of a neurenteric cyst dorsal to the brain stem extending into the fourth ventricle. Although the cyst had an unusual location, what makes this case truly informative is the visual deficit that the patient experienced after surgery. The patient complained of double vision developing in the early perioperative period, and formal examination by a neuro-ophthamologist diagnosed the patient with a one and a half syndrome. This syndrome is typically related with damage to the paramedian pontine reticular formation (PPRF) and the medial longitudinal fasciculus (MLF) on the same side. Since neither of these structures was within the area of the cysts, it is puzzling as to why they would have been affected by the cysts' removal, as one and a half syndrome is typically caused by intrinsic brain stem or pontine lesions such as tumor or stroke [15].

Since the postoperative MRI did not reveal bleeding or ischemia of the brainstem, the authors hypothesize that in the process of cyst removal, a midline raphe became disturbed. The vibrations of these fibers, upon the removal of the cysts, resulted in damage more deeply within the pons in the PPRF and the MLF. This hypothesis is supported by the observance of abnormal midline raphe during the second surgery and supports the notion of the "split notochord" hypothesis. This hypothesis may explain the persistence of endodermal element dorsally in the fourth ventricle without an anterior cyst component or persistent of a cleft in the anterior brainstem which connects the cells to their endodermal origins.

Conclusion

Neurenteric cysts are rarely identified in the fourth ventricle of the brain, as they are a developmental cyst originating from the primitive connections between the nervous system and the endoderm of the gastrointestinal system. Given this origin, it follows that a neurenteric cyst occupying the fourth ventricle would have an intervening connection to the ventral portions of the nervous system. In this case report, a midline raphe was identified. It is speculated that traction to this midline raphe during removal of the cysts led to transmission of a vibrational or traction injury to the pons resulting in a one-and-a-half syndrome. Although both neurenteric cysts and ophthalmoplegic complications of dorsal fourth ventricular surgery are rare, care must be taken in removing dorsal cysts so as not to distort or place traction upon possible connections between the cyst and the brainstem itself.

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