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Rapid Visual Deterioration Caused by Posterior Fossa Arachnoid Cyst in an Adolescent: A Case Report and Review of the Literature

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Abstract

Posterior fossa arachnoid cysts are rare posterior fossa lesions that are considered to be mostly congenital in origin. Generally, these lesions are benign, asymptomatic and are found incidentally in most cases. In some rare cases, they can be symptomatic and are diagnosed in childhood or adolescence. Here, we described a case of posterior fossa arachnoid cyst in a 16-year-old girl, admitted for rapid visual deterioration following 6 months of high intracranial pressure with cerebellar ataxia.

Keywords

Posterior Fossa, Arachnoid Cyst, MRI, Surgery

1. Introduction

Intracranial arachnoid cysts are rare benign extra-axial lesions corresponding approximately to 1% - 2% of all intracranial masses [1] [2]. They had been described by Richard Bright in 1827 and were named "serous cysts" [3]. These lesions occurred within the split layers of the arachnoid and are filled with Cerebrospinal Fluid (CSF) and are considered to be mostly congenital in origin. They are uncommon in the posterior cranial fossa with only 5% - 48% of arachnoid cysts located in this area from which retro cerebellar locations are most represented [4] [5]. They are usually incidental findings in asymptomatic patients [6]. In some rare cases, these cysts can be enlarged and become symptomatic. Here, we report a rare case of Posterior Fossa Arachnoid Cyst (PFAC) revealed by ataxia and rapid

visual deterioration before admission.

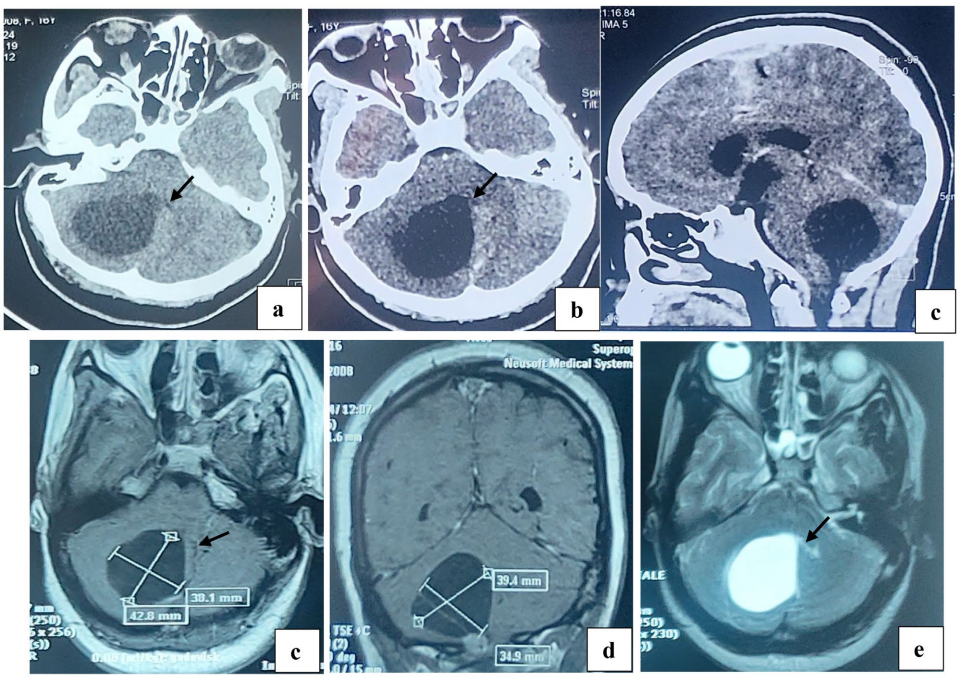
2. Case Report

A 16-year-old girl with no previous medical history was admitted to our emergency room for abrupt visual deterioration for 02 days. In interrogatory, this girl complained of headache, vomiting and gait disturbance for 06 months. Ophthalmologist test found bilateral papilledema and decreased visual acuity (1/10 in both eyes). Neurological examination hadn't noted any abnormalities apart from cerebellar ataxia. Brain CT scan revealed a huge posterior fossa cyst compressing the 4th ventricle and the surrounding cerebellum and causing hydrocephalus. This mass was intracerebellar, well-rounded, with a thin hypodense capsule that did not enhance after contrast administration. According to the clinical signs and the proximity of this cyst to the 4th ventricle, we recommended a brain MRI to best explore this lesion. MRI showed a huge intracerebellar cyst filling the right posterior fossa measuring 42.8 mm × 38.1 mm × 34.9 mm, compressing the 4th ventricle and confirming the CT scanning data and had the same characteristics of CSF fluid in different sequences (**Figure 1**). The diagnosis of PFAC was strongly suggested. In prone position and after general anesthesia, a median suboccipital craniectomy was done through a midline incision following a "Y" shaped durotomy. After a puncture with an intravenous cannula G 18, a small cerebellar corticectomy was performed to remove all the cyst wall. A histopathological study of excised tissues stained with hematoxylin and eosin showed arachnoid mesothelial cells arranged among fibers of the connective tissue confirming our diagnosis. After surgery, headache and papilledema disappeared in 02 weeks and ataxia improved in six months. Postoperative CT scan was done 1 week later and repeated 1-year follow-up showed no recurrence. Our patient is free of symptoms and has completely recovered her visual acuity (10/10 in both eyes) (**Figure 2**).

3. Discussion

An arachnoid cyst consists of a cavity lined with arachnoid cells and filled with fluid that closely resembles CSF [7]. The posterior fossa is the second most common location of arachnoid cysts after the middle cranial fossa [8] [9]. Most of them are clinically silent and remain static in size; however, on rare occasions, these cysts can increase in size and produce symptoms due to mass effects [10]-[12]. Several mechanisms could be responsible for gradual enlargement of an AC: intracystic hemorrhage, an osmotic gradient allowing a passive fluid diffusion into the cyst, a ball-valve mechanism, or an active secretion from the cyst wall are the main theories to explain cyst growth [5] [13]. The clinical signs vary depending on the age, the lesion size and degree of compression in adjacent structures in the posterior fossa. No specific signs such as headache, vomiting, visual deterioration, gait disturbance or ataxia, dizziness, hearing loss, tinnitus, lower cranial nerve palsies, facial numbness, cerebellar and pyramidal signs, psychomotor retardation, seizures could be seen [13] [14]. In our case, headache, vomiting and ataxia were

the most clinical symptoms. Visual deterioration had been added just 03 days before admission and was caused by acute hydrocephalus. This visual symptom had been cited by Shin C. J. *et al.* and can be explained by the increased Intracranial Pressure (ICP) due to PFAC compression of the 4th ventricle and disruption of the CSF flows [15]. The best radiological investigation tool is MRI to explore PFACs. This exam permits the distinction of PFACs from other cystic formations in adulthood such as cerebellar cystic astrocytoma, cystic hemanglioblastoma, epidermoid or dermoid tumors [16]. In an emergency, we did the CT scan first and put the patient on intravenous steroids regimen (Methylprednisolone 120 mg per day) just in time to have the MRI. The treatment of choice for most cases of ACs is conservative. In cases of progressive neurological deficit or persistent symptoms, surgery is indicated. The objective of the surgery is to collapse the cyst and improve the clinical manifestations [2] [17]. Treatment protocols vary, including craniotomy for cyst excision, craniotomy for fenestration, endoscopy for fenestration, or cystoperitoneal shunting [2] [14] [17]. Surgical series for arachnoid cysts demonstrate the improvement of symptoms in approximately 80% of patients and complications occur in approximately 10% - 15% of cases [18] [19]. Microsurgical fenestration via open craniotomy provides direct access to the cyst and allows partial or complete excision of the cyst membrane and is often indicated for large or multiloculated cysts [20]. By this procedure and depending on surgeon expertise, postsurgical recurrence rate is very low in such cases like ours.



Preoperative MRI showed the same PFAC with the same characteristics of CSF fluid in different sequences (T1W images **cd** and T2W images **e**).

Figure 1. Preoperative CT scan before and after contrast administration in axial view (a), (b) and sagittal view (c) showing a large PF arachnoid cyst compressing the fourth ventricle (black arrow) and adjacent structures.

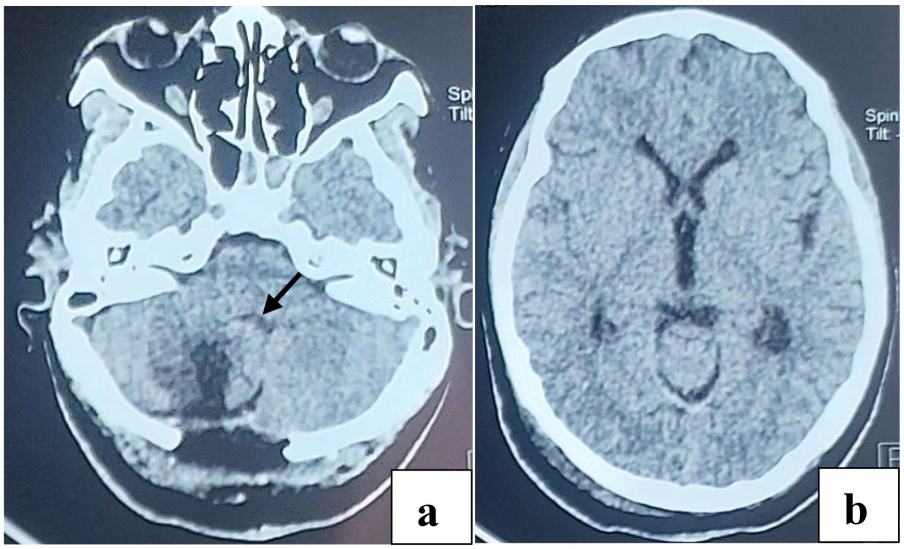


Figure 2. (a) and (b) Postoperative views showing no residual mass and apparition and normalization of the 4th ventricle (black arrow) and the other ventricular cavities.

4. Conclusion

Posterior fossa arachnoid cyst is a rare event in neurosurgical practice. Clinical symptoms related to rapid visual deterioration are very rare. In such a case, urgent excision of the cyst must be done without delay.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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