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Intracranial ependymal cyst with unusual presentation: Case report and review of literature☆

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ABSTRACT

Background: Ependymal cysts remain a very rare cause of neurological symptoms among intracranial cystic lesions. It differs from an arachnoid cyst, as it does not communicate with subarachnoid space.
Case description: A 33-year-old woman was admitted in our neurosurgery department with history of headache, vomiting and right unilateral blindness. Neuroimaging studies showed an intraparenchymal cystic lesion in the right frontal lobe with radiologic characteristic as seen in the ependymal cyst or hydatic cyst. She underwent the neurosurgical procedure of marsupialization and was discharged on day 6 post op. At one month follow up, the patient presented with a decrease vision acuity on the left eye, and head CT scan showed a recurrence of the cyst hence, we performed a second surgery with shunt procedure. Histological examination of the resected tissue demonstrated that, it was an ependymal cyst.
Conclusion: Since there is often times a clinico-radiological discordance they are generally incidental findings in majority of cases. While complete surgical excision is the treatment of choice for the symptomatic cysts, cystic fluid diversion presents a better compromise nevertheless in cases where this may not be achieved.

1. Introduction

Ependymal cysts represent benign congenital ependymal-lined cysts [1,2] and constitute a very rare cause of neurological symptoms among intracranial cystic lesions [3,4]. As term of terminology, they are designed by several synonyms as “ependymal cyst”, “neuroepithelial cyst”, “glioependymal cyst” and neuroglial cyst” testifying the difficulty of naming this disorder. They are generally located inside the lateral ventricles or in the juxtaventricular regions, particularly the temporoparietal and frontal lobe [2]. They are considered to originate from sequestration of developing neuroectoderm during embryogenesis [2,5]. The cysts often occur intracranially in the cerebral parenchyma; however, some cases have been described elsewhere in the cerebral nervous system such as posterior cerebral fossa and spinal cord [6,7].

In this article, we report the case of an intracranial ependymal cyst in a young woman with an unexplained blindness.

2. Case report

2.1. History and examination

A 33-year-old woman was referred to the department of neurosurgery of Fez, Morocco by an ophthalmologist with a history of blindness. She came from the surrounding countryside with a good socioeconomic background and no reported contact with animals like dogs or other carnivores whatsoever.

She had experienced headaches, intermittent episodes of vomiting for about 3 years' period and consulted only when her vision reduced especially in the right eye approximately a year before her admission in our department. The neurological examination revealed no motor weakness nor sensory disturbances and the deep tendon reflexes were equally normal. The cerebellar functions were intact. However, the ophthalmological examination revealed light perception in the right eye with a large slowly reacting pupil while the acuity was normal in the contralateral left eye 10/10. Fundoscopy revealed bilateral

Abbreviations: CSF, cerebrospinal fluid; CT, computerized tomography; MRI, magnetic resonance imaging

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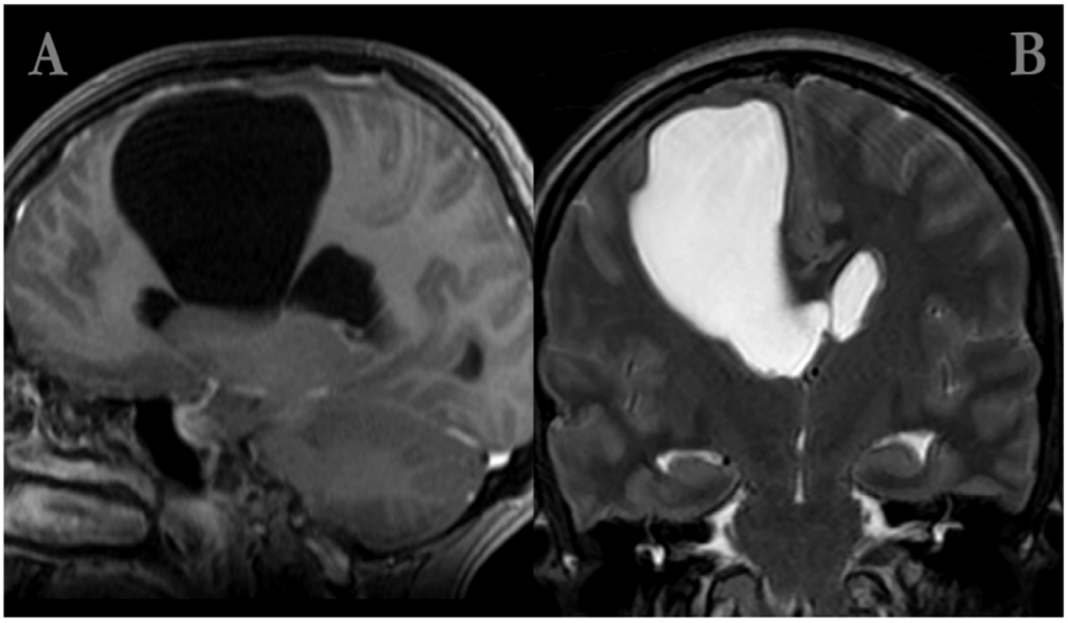

Fig. 1. Initial MRI in T1 weighted sagittal (A) and T2 weighted coronal view showing a large cystic lesion in the frontal lobe with mass effect on the lateral ventricle and adjacent structures. The signal of the lesion is similar to that of CSF, with no enhancement after gadolinium.

papilledema. The ocular motility, anterior segment and ocular tonus were normal.

2.2. Neuroimaging

MRI revealed a large cystic lesion in the right frontal lobe. It was well circumscribed, 3 × 5 cm in diameters, and have had a signal parallel to that of the cerebrospinal fluid (CSF) in both T1 and T2 weighted sequences (Fig. 1).

2.3. Operation

The patient underwent two neurosurgical procedures. Firstly, the cyst has been approached and was excised through a small right frontal corticotomy. The lesion appeared to be cystic mass with thin membrane, containing watery fluid, CSF-like and its wall was not clearly separated from the brain parenchyma. During surgery, a fenestration of the cyst to the lateral ventricle was done with subtotal removal of the cyst wall.

2.4. Postoperative follow-up

In the immediate postoperative period, an epileptic seizure occurred and was controlled by antiepileptic drug. Postoperative brain CT scan showed a good opening of the cyst to the lateral ventricle and a significant reduction of its volume (Fig. 2). The biochemical lab analysis of cystic fluid showed protein (0.14 g/l) and glucose (0.60 g/l). The patient recovered slightly from her initial symptoms in such a way that she was free from signs of increased intracranial pressure and gained some vision in her right eye.

At follow-up, the patient complained reduced vision on the left eye with vision acuity of 2/10, papilledema and dyschromatopsia. Brain CT scan at follow-up showed a recurrence of the cyst with a noticeable midline shift (6.73 mm) due to mass effect. Then, a cystoperitoneal shunt procedure was done with diversion of the cyst content to the peritoneal cavity. Postoperatively, her vision significantly improved and the patient remained symptom-free at a mean follow-up of one year.

2.5. Pathological findings

Histological examination of the cyst wall revealed a lining epithelium composed of simple cuboidal epithelial cells (Fig. 3). The epithelial cells showed diffuse positivity for glial fibrillary acidic protein. So, the histological diagnosis was an ependymal cyst.

3. Discussion

Intracranial ependymal cysts are rare lesions that occur less frequently than arachnoid cysts [1,3,4]. As stated by Friede and Yasargil [8], Zehnder was the first author to describe this entity in 1938. Currently, our search on PubMed find approximately only 39 cases reported in literature. Table 1 presents the 24 cases in adult reported [1,3,6–12] on the intracranial ependymal cysts since the literature review of Friede and Yasargil who collected 15 cases in 1977; we excluded the cases from congenital or neonates because of the ambiguity of the diagnostic with congenital subependymal pseudocyst. Typically, ependymal cysts are neuroepithelial cysts that arise from ectopic rests of primitive neuroglial tissue, and hence, can arise anywhere in the neuraxis but are often located in the central white matter of the temporoparietal and frontal lobes in juxtaventricular position [2,5,7,12].

The ependymal cysts seem to occur at young age within the second and third decades [12]. However, some cases have been reported at all ages including newborn and children [1,13,14] and hold the older population [7]. There is not a consensus about the sex predilection. However, our search found a female predominance in the older population and indeed vice versa in the younger age group as we see the cases reported in the literature. Indeed, El Damaty et al. [7] described six cases including five females with age ranged from 42 to 67 years old. Conrad et al. [6] described eight cases with seven females from 35 to 60 years old. In contrast, Makhoul et al. [15] reported 10 children with only 3 females. Our patient has just turned 33 years old and was female.

The clinical presentation consists of a spectrum of symptoms and range from being asymptomatic, to seizure episodes and indeed focal and lateralizing signs. Visual field defects have also been documented and other non-specific signs. Harada et al. [4] reported a case with an unusual presentation with hemifacial spasms. Our patient had reduced vision and the clinical examination revealed a visual field defect only on the right side of which we did not find a link with the intracranial

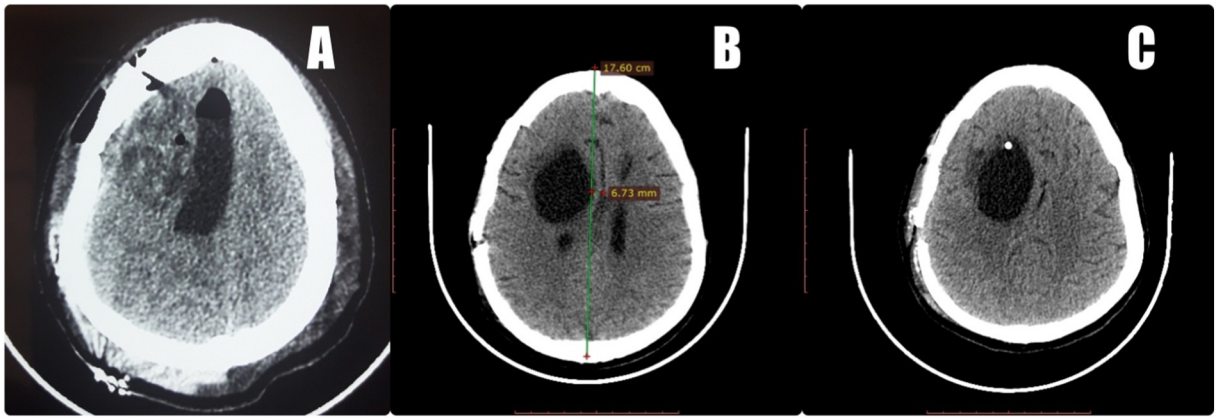

Fig. 2. (A) Immediate axial section postoperative CT scan showing a good marsupialization of the cyst to the lateral ventricle; (b) two months' follow up CT scan showing a mass effect with midline shift and (c) ain CT scan of control after shunt procedure (B).

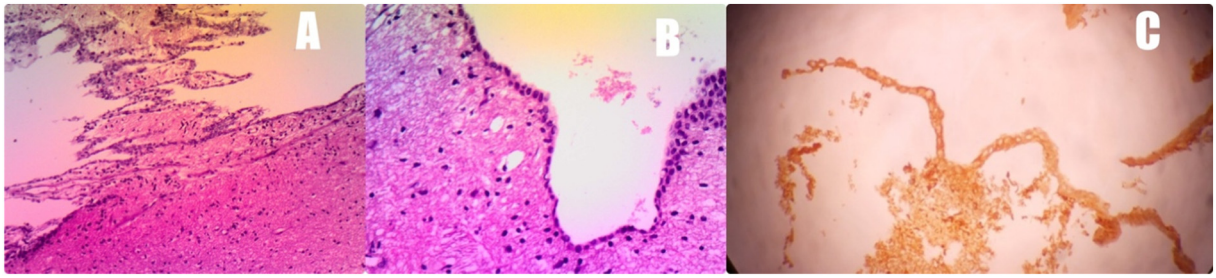

Fig. 3. Histological examination of the cyst wall showing epithelium composed by simple cuboidal epithelial cells without prominent cilia [Hematoxylin & eosin, ×100, (A) ×200 (B)] and positive GFAP staining.

Table 1
Cases reported on intracranial ependymal cysts since the literature review[a] of Friede & Yasargil in 1977.

Authors (year)	Cases	Age	Sex	Location
Friede & Yasargil 1977	1	62	F	Left frontal
Friede & Yasargil 1977	2	37	M	Left frontal
Ho & Chason 1987	3	30	M	Cerebellopontine, subarachnoid
Tsuchidas et al. 1997	4	29	M	Cerebellum
Boockvar et al. 2000	5	43	M	Lateral ventricle
Kanazawa et al. 2000	6	17	M	Pineal region
Pawar et al. 2001	7	14	M	Temporal horn of lateral ventricle
Frazier et al. 2004	8	55	F	Left cerebellum
Conrad et al. 2008	9	35	F	Tectum of the mesencephalon
Conrad et al. 2008	10	53	F	Mesencephalon, pons, left thalamus
Conrad et al. 2008	11	54	F	Mesencephalon
Conrad et al. 2008	12	56	F	Mesencephalon
Conrad et al. 2008	13	60	F	Mesencephalon, thalamus
Conrad et al. 2008	14	35	F	Right mesencephalon, thalamus
Conrad et al. 2008	15	36	M	Left mesencephalon/thalamus
Conrad et al. 2008	16	22	F	Mesencephalon
El Damaty et al. 2017	17	67	F	Right parietal
El Damaty et al. 2017	18	67	F	Right occipital
El Damaty et al. 2017	19	42	F	Midbrain
El Damaty et al. 2017	20	60	F	Left occipital
El Damaty et al. 2017	21	61	M	Right parietal
El Damaty et al. 2017	22	65	F	Left frontoparietal
Umredkar et al. 2010	23	25	F	Right frontal
Umredkar et al. 2010	24	25	M	Left temporal

[a] Friede and Yasargil collected 15 cases described in the literature in 1977 and reported 2 own cases. Adding theses 24 reported cases, it becomes 39 cases.

ependymal cyst.

In terms of radiologic features, ependymal cysts have the same signal as CSF. "The best diagnostic clue is a non-enhancing thin-walled CSF-containing cyst of the lateral ventricle" [2]. The differential diagnoses include arachnoid cyst from which it differs as it does not often show any communication with the arachnoid space, the choroid plexus cyst and asymmetrical ventricle. Of importance is our North African setting where hydatid disease is endemic; the ependymal cysts closely resemble a simple brain hydatic cyst. While the anamnesis to point to socioeconomic context and contact with dogs may help in the probability of one diagnosis over the other the neuroradiological characteristics by both the CT and MRI are quite nonspecific to separate the two, hence to keep an open mind in the surgical approach. The role of other diagnostic tests to help to differentiate the two diagnosis can be explored [1], but is not the interest of this work.

There is no consensus regarding the surgical approaches for ependymal cysts. Neurosurgical intervention is necessary for symptomatic ependymal cysts. However, in case of asymptomatic patient, the neurosurgical intervention should be seriously considered [1,12,16] and a case by case consideration discussion is advised. Many approaches were described to surgically treat the ependymal cysts. A subtotal resection of the cyst wall with marsupialization was performed in our case and during the second surgery, we performed a cystoperitoneal shunt because of the early recurrence of the cyst. Pawar et al. [1] and Tanje et al. [5] suggested that total resection of the cyst is the ideal option even though this will not be possible in cases of the adherence of the cyst wall to the adjacent brain parenchyma with no clear cleavage plane. For those cases, a simply marsupialization with near-total resection of the lateral wall of the cyst should be performed and a cystoperitoneal shunt may be helpful to prevent a recurrence.

4. Conclusion

Ependymal cysts are rare congenital cause of neurological deficits among intracranial cysts. It occurs mostly in the supratentorial compartment. Since there is often times a clinico-radiological discordance they are generally incidental findings in majority of cases. While complete surgical excision is the treatment of choice for the

symptomatic cysts, cystic fluid diversion presents a better compromise nevertheless in cases where this may not be achieved.

Disclosures

"We do not have any financial or organizational relationships with commercial interests or others entities. We hereby certify that to the best of our knowledge, no aspect of our current personal or professional circumstances places us in the position of having a conflict of interest with our duties, responsibilities, and exercise of independent judgement."

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