

EPENDYMAL BRAIN CYST

İzzet Övül M.D., Kazım Öner M.D., Özcan Binatlı M.D. and Eren Demirtaş M.D.

Departments of Neurosurgery and Pathology (ED) University of Ege (Aegean) 35100 BORNOVA - İZMİR

Turkish Neurosurgery 1 : 49 - 51, 1989

SUMMARY :

A case of subarachnoid ependymal (epithelial) cyst is described. It was located in the frontoparietal lobe of the brain, was lined with cuboidal epithelial cells, and had no communication with the ventricular system. The only clinical sign was epilepsy. Diagnosis was demonstrated by computerized tomography (CT). The cyst was removed and communication was created with the subarachnoid space. This rare pathology is discussed together with a review of the literature.

KEY WORDS :

Ependyma. Epithelial cyst. Intracerebral cyst.

INTRODUCTION

Ependymal cysts lacking communication with the cerebrospinal fluid (CSF) pathways have been observed throughout the nervous system (12). Supratentorial ependymal cysts are rare entities that may occur in either an intracerebral or a subarachnoid location (1,2,3,5,6,8,9,10,13,14,20). In 1979, Markwalder and Zimmermann reviewed 21 reported cases of intracerebral or convexity ependymal-lined cysts without communication with the subarachnoid space, and added one case of their own. There have been 3 additional cases, 1 reported by Haddad et al. (7) in 1982, 1 reported by Pearl et al. (15) in 1982 and 1 reported by Oro et al. (12) in 1983, bringing the total number to date to 24. We present an additional case of an ependymal cyst, and review the previously reported cases of supratentorial ependymal cysts with emphasis on treatment and results.

CASE REPORT.

A 25 year-old man was admitted to our clinic with a 6-month history of headache and generalised epilepsy. There was no previous history of significant head trauma, infection or other central nervous system (CNS) disorder.

The general physical examination was unremarkable. A neurological examination revealed no abnormalities of cranial nerve function, and sensory, motor, cerebellar, and reflex testing were normal. There was no evidence of papilledema. Laboratory investigations revealed no abnormality. Serological tests for hydatid disease were normal. Skull and chest x-ray films were uneventful. EEG was normal. Ultrasonography of the abdomen was normal. A Computed tomography (CT) scan revealed a

left frontoparietal, non-enhancing extra axial cyst (fig 1).

A left frontoparietal craniotomy was performed. The duramater over the left cerebral hemisphere was tense and fluctuating on palpation. When the dura was opened, a thin walled cyst ruptured. The cyst wall presented on the cortical surface in the left frontoparietal region. It consisted of a thin translucent membrane, continuous with the pia arachnoid but not communicating with the subarachnoid space. The cyst contained CSF-like clear fluid and compressed the surrounding brain tissue. Cyst fluid was aspirated and the cyst was resected, subtotally, creating free communication with the subarachnoid space.

The postoperative period was uneventful. At examination 8 months after surgery, he was doing well.

Light microscopic examination of the cyst wall revealed a layer of loose collagenous tissue lined by a single layer of cuboidal epithelial-ependymal cells. In some areas, however, the cells had a pseudostratified appearance. No cilia were identified (fig 2-3).

DISCUSSION

Ependymal cysts related to the subarachnoid space have been reported in the parasagittal region, about the Sylvian fissure and in the basal subarachnoid cisterns and posterior fossa (2,9,14,20). Intracerebral examples have been found chiefly in the cerebral hemispheres, although they have also been reported in the thalamus, quadrigeminal plate, and cerebellar vermis (2,4,9,10,14,20). Analysis of these



Fig 1. Contrast enhanced CT scan. A non-enhancing low density mass lesion is present in the left frontoparietal region. The density of the lesion resembles that of cerebrospinal fluid. The shape, smooth contour, and density of the mass imply a fluid-filled cyst.

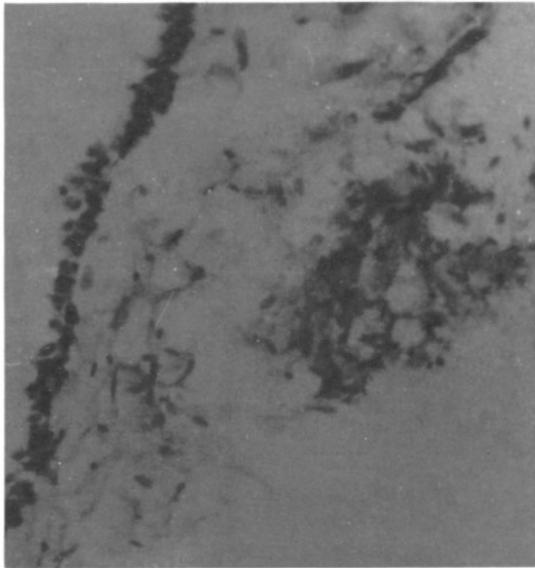


Fig 2 : Part of cyst wall lined by pseudostratified cuboidal ependymal lying upon loose connective tissue. H. and E.X440.

cases reveals that they appear in adult life (2,3,5,6,8,9,10,13,14,20). The youngest case was a four-month-old infant reported by Haddad et al, in 1982 (7). They generally occur in females, with a 3:1 preference, and are most commonly located in the frontal or frontoparietal region (15). The most common presenting symptom is seizures, followed by headache and loss of consciousness with the clinical

history generally lasting less than 2 years before diagnosis. The cysts are large, most being at least 4 cm. in diameter. The fluid in the cysts was clear in nearly half of the reported cases. In others, it was opalescent, milky or turbid. The lining consisted of flattened to cuboidal or columnar cells. In approximately half of the cases apical cilia have been observed.

The origin of epithelial cysts related to the central nervous system is still debated. It is generally thought that they result from a developmental defect, although the mechanism of origin is unknown. One theory holds that the cysts arise from ependymal rests that have become displaced into the cerebral substance and subarachnoid space during embryonal development. Displacement may have been caused by malformation of the mantle layer or by punching off a diverticulum resulting in an isolated ependymal cavity (2,3,6,8,9,14,20) or by differentiation spongioblastic cells into ependyma about a primary encephaloclastic intracerebral defect occurring in early faetal life (17). Heterotopic glial nests, some containing ependymal lined canals, are reported defects of the central nervous system (16). Friede and Yaşargil prefer the theory of displacement of a segment of the wall of the neural tube into the brain substance or subarachnoid space with consecutive development of a pure intracerebral cyst or a cyst which maintains its communication with the CSF - space (5). An alternate hypothesis, however, is that the different forms of cysts may be of differing origin, arising from heterotopic or embryologically displaced epithelium, as has been proposed for all "neuroepithelial" cysts (19). The clinical course suggests that these cyst expand, although the cause of expansion is unknown. Jakubiak and co-workers suggested that the hyperosmolality of the cyst fluid draws water from the surrounding tissue into the cyst causing it to enlarge; the four-fold elevation of protein in the contents of the cysts was used as evidence (8). However it is difficult to explain why the protein level should rise in later life, and if it is high from the onset, why it does not draw in fluid early in life. Meanwhile, in some cases cyst fluid does not have elevated protein. Active secretion of fluid into the cyst by the epithelial cell lining is another possibility (5,6,8). However, it does not explain why the secretion should occur many years after the assumed formation of the cyst.

Our case seems to resemble the 24 supratentorial neuroepithelial cysts reported in the literature. There seems to be a close relationship to the cases reported by Tandon et al., Ghatak et al, and Friede et al. (5,6,20).

From the therapeutic point of view, various treatments have been suggested. These have included repeated aspiration, partial or if it is possible, total excision, and shunting (2,5,9,13,14,17,19,20). The goal

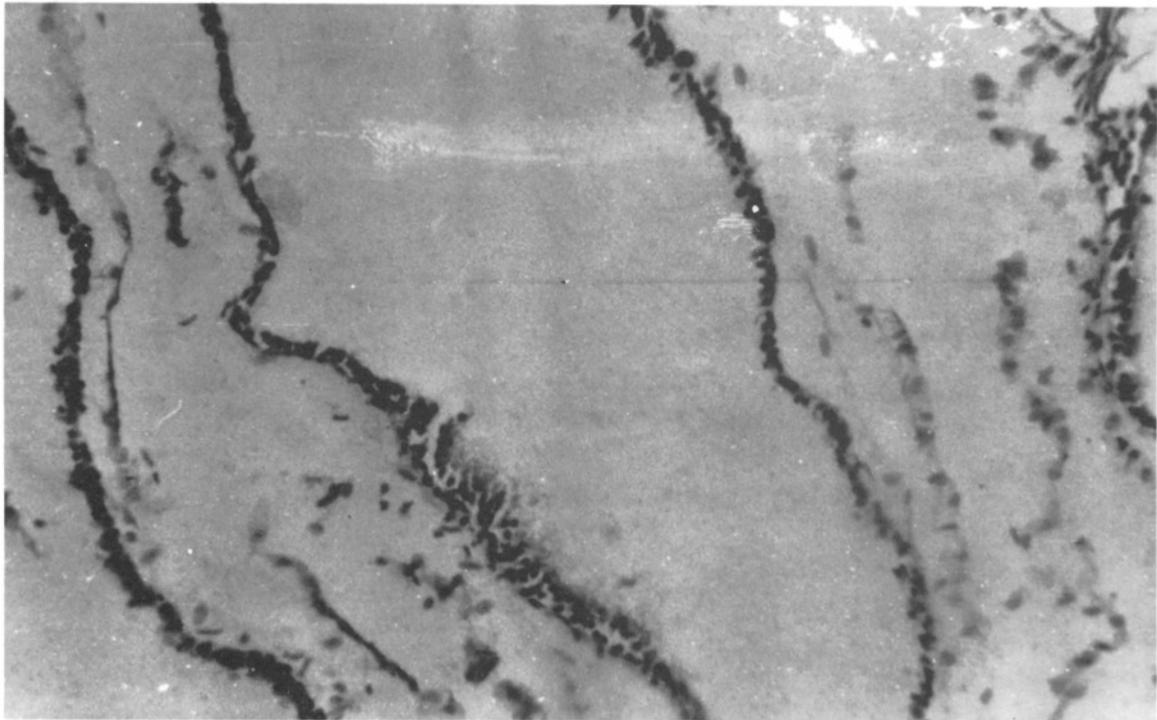


Fig 3. Another section of zone the cyst wall is showing cuboidal epithelium and loose connective tissue.

of treatment is to provide adequate communication of the cyst with the CSF pathways. Aspiration is not practical because of the need for repeated therapy. Total excision of the cyst is often not possible. The majority of supratentorial lesions have been treated with subtotal resection unroofing the cyst to create communication with either the ventricles or the subarachnoid space (4,5,10,14,20). Although reepithelisation may occur after partial resection, it is infrequent (9,10,11,18). In our case the cyst wall was resected subtotally and created communication with the CSF space. Various shunting procedures with or without cyst wall resection have also been used (15,18).

REFERENCES

1. Aicardi J, Bauman F: Supratentorial extracerebral cysts in infants and children. *J Neurol Neurosurg Psychiatry* 38:57-68, 1975
2. Bhandari YS: Noncommunicating supratentorial cysts. *J Neurol Neurosurg Psychiatry* 35:763-770, 1972
3. Bouch DC, Mitchell I, Maloney AES: Ependymal lines paraventricular cerebral cysts: a report of three cases. *Journal of Neurology Neurosurgery and Psychiatry* 36:611-617, 1973
4. Davis RP, Sachdev VP: Ependymal cyst of the quadrigeminal plate. Case report. *Neurosurgery* 16:96-99, 1985
5. Friede RL, Yaşargil MG: Supratentorial intracerebral epithelial (ependymal) cysts: review, case report, and fine structure. *Journal of Neurology, Neurosurgery and Psychiatry* 40:127-137, 1977
6. Ghatak NR, Itirano A, Kasoff SS, Zimmerman HM: Fine structure of an intracerebral epithelial cyst. *J Neurosurg* 41:75-82, 1974
7. Haddad FS, Abla A, Allam C: Ependymal brain cyst. *Surg Neurol* 18:246-249, 1982
8. Jakubiak P, Dunsmore RH, Beckett RSV: Supratentorial brain cysts. *Jeurosurg* 28:129-136, 1968
9. List CF, Williams JR: Subdural epithelial cyst in the interhemispherical fissure: Report of a case with some remarks concerning the classification of intracranial epithelial cysts. *J Neurosurg* 18:690-693, 1961
10. Macgregor BJL, Gawler J, South JR: Intracerebral epithelial cysts. *J Neurosurg* 44:195-198, 1979
11. Markwalder TM, Zimmerman A: Intracerebral ciliated epithelial cyst. *Surg Neurol* 11:195-198, 1979
12. Oro JJ, Ansbacher LE, Levy WS: Septate bifrontal ependymal cyst: Case report *J Neurosurg* 59:344-347, 1983
13. Palma L: Supratentorial neuroepithelial cysts. Report of two cases *J Neurosurg* 42:353-358, 1975
14. Patrick ES: Ependymal cyst of the Sylvian fissure: case report *J Neurosurg* 35:751-754, 1971
15. Pearl GS, Takei Y, Tindall GT, et al: Supratentorial subarachnoid epithelial cyst: Case report and ultrastructural study. *Neurosurgery* 11:48-51, 1982
16. Papoff N, Feigin I: Heterotopic central nervous tissue in subarachnoid space. *Arch Pathol* 78:533-539, 1964
17. Rand BO, Foltz EL, Alvord EC Jr.: Intracranial telecephalic meningoencephalocele containing choroid plexus. *J Neuropathol Exp Neurol* 23:293-305, 1964
18. Rilliet B, Berney J: Benign ependymal cyst of pons. *Childs Brain* 8:1-8, 1981
19. Shaughshoti S, Netsky MG: Neuroepithelial (colloid) cysts of the nervous system: Further observations on pathogenesis, location, incidence and histochemistry. *Neurology (N.Y.)* 16:887-903, 1966
20. Tandon PN, Roy S, Elvidge A: Subarachnoid ependymal cyst. *J Neurosurg* 37:741-745, 1972