Ependymal cyst of the Sylvian fissure

Case report

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An ependymal cyst occurring in the Sylvian fissure is described. No ependymal cysts have been reported above the tentorium and external to the cerebrum. Concepts of the origin of ectopic ependymal tissue and other glial heterotopias are discussed.

KEY WORDS ependymal cyst Sylvian fissure heterotopia hamartoma

Cysts occurring in and about the lateral, third, and fourth ventricles, having linings consisting of a single layer of cuboidal epithelium, have been called "choroid plexus cysts," "ependymal cysts," or "epithelial cysts," while certain of those of the third ventricle have been designated "colloid" or "paraphysial cysts." Those occurring within or in connection with the ventricular system are relatively common; however, ependyma-lined cysts that are not contiguous with the ventricular system or central canal of the spinal cord are quite rare. There are no previous reports of such cysts occurring above the tentorium, external to the cerebrum. This report concerns such a cyst occurring in the Sylvian fissure.

Case Report

A 58-year-old housewife was admitted in October, 1958, because of progressively severe headache, diplopia, and a feeling of dizziness. She described recurrent right temporal headaches for the previous 5 or 6 years, occasionally accompanied by nausea and vomiting. For the past 6 to 12 months she had complained of occasional transient diplopia and occasional dizziness during which she noted a tendency to stagger or deviate her direction when walking. She did not recall whether her deviation was to the right or to the left. During the recent 2 to 3 months, she had noted a very occasional transient numbness in the left hand but denied weakness or persisting disturbance of function of that hand.

This history was confirmed by her family with an additional history of 6 to 12 months of progressive personality change manifested by increased irritability, lack of attention to personal care, a tendency to ramble during conversation along with some inappropriate affect.

Examination. There were no definitive neurological abnormalities except for generally hyperreactive reflexes with some minimal weakness in the left hand. There was no papilledema. A lumbar puncture revealed a pressure of 140 mm H2O and normal spinal fluid protein. Skull films were normal. An angiogram revealed a marked medial shift of the right middle cerebral arterial group, indicating a mass in the right Sylvian fissure (Fig. 1).
Operation. On October 24, a right temporal trephine opening was made. Immediately beneath the dura a cyst was encountered enclosed by a fine transparent membrane. The trephine opening was enlarged to a small temporal craniectomy. The cyst contained colorless, water-clear fluid and was drained. On the assumption that this was a simple arachnoid cyst, no fluid specimen was taken. The underlying cerebral cortex in the midportion of the Sylvian fissure had been displaced medially approximately 4 cm. The insula, however, was not exposed. The cortex, other than having somewhat flattened gyri, appeared to be normal. A few fine blood vessels ran through the transparent membranous wall of the cyst. There was no apparent communication with the adjacent subarachnoid space nor with the ventricular system. The cerebral cortex was displaced in a manner similar to that seen with meningiomas arising from the dura mater overlying the Sylvian fissure, suggesting that this cyst also arose superficial to, rather than within, the Sylvian fissure. There was no evidence of cortical undermining or subcortical or intracerebral expansion of any portion of this cyst. The volume of the emptied cyst was 60 cc.

A few bits of tissue membrane were removed for routine pathological examination. The cavity was filled with air, and postoperative x-ray films were made (Fig. 2).

Microscopic examination of the cyst wall revealed low cuboidal epithelium made up of a single layer of cells having fairly large centrally positioned nuclei. The cells were characteristic of ependyma but without cilia (Fig. 3).

Postoperative Course. The patient recovered without complications. All symptoms including the personality disturbance disappeared. She would not return for follow-up examination but recent communication with the patient 12 years following...
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surgery indicates that there was no residual symptomatology referable to the head or brain.

Discussion

Extramedullary ependymal cyst has been reported in the cervical spinal canal by Hyman, et al., 4 as well as by Moore and Book. 5 Both reports concerned cysts that were clearly extramedullary. Dandy, 2 in his monograph Surgery of the Brain, reported an ependymal cyst of the cerebellopontine angle which he felt had arisen from a continuation of the choroid plexus of the lateral foramen of Luschka. Hamby and Gardner 3 reported an ependymal cyst in the region of the corpora quadrigemina in a 16-year-old girl. This cyst apparently did not communicate with the ventricular system. Hamby hypothesized that the isolated island of tissue might have evaginated from the posterior third ventricle. Vitek, et al., 8 reported an ependymal cyst of the thalamus, probably of similar origin.

In our case, this hypothesis is less applicable because the cyst was not close to the ventricular system. Nonetheless, even in this case the concept of an isolated nest of ependymal cells is at present the most acceptable explanation.

Russell and Rubenstein 6 suggest that the element of maldevelopment with segregation of either immature or mature cells is essential to the interpretation of many forms of tumor found in the central nervous system. They suggest that this segregation occurs during embryonic or early fetal life but that nevertheless the cells may go on to attain cytological maturity. They agree that these heterotopias share with other tissue the possibility of becoming neoplastic but concluded that their neoplastic potential was not greater than normal tissues. Cooper and Kernohan, 1 on the other hand, concluded that these heterotopic cell nests developed as a result of either pinching off of glia, or of glia streaming through pial defects, or by direct invasion of the pia; they suggested that these heterotopias often appear at a much later stage of fetal development, and even after maturity. Yakovlev and Wadsworth 9,10 in discussing schizencephalies (true clefts formed in the brain as the result of a failure of development of the cerebral mantle in the zones of cleavage of the primary cerebral fissures) emphasized the embryonal origin of these symmetrical clefts, heterotopia of cerebral substance in the subarachnoid space, and other dysgenetic anomalies. They concluded that these disorders were essentially agenetic processes occurring as a result of more general factors acting upon the fetal nervous system not later than the second month of gestation.

The frequent occurrence of ependymal cells in subarachnoid glial heterotopias 1 and the high incidence of ependymomas apparently developing from these heterotopias are additional factors supporting the thesis of embryonal maldevelopment as the most likely mode of origin of neuroglial heterotopias.

Summary

A case of ependymal cyst overlying the Sylvian fissure is described. It is unique in that no other ependymal cyst has been reported occurring external to the brain above the tentorium. The patient is alive and well 12 years following surgery, with no evidence of recurrence.
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