Leptomeningeal cyst of the posterior fossa

Case report

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A case of a posterior fossa extradural cyst is presented in which both the cyst and the arachnoiditis that accompanied it were thought to have been caused by trauma which had occurred 16 years earlier. The reasons why this was diagnosed as a leptomeningeal cyst are discussed.

KEY WORDS · leptomeningeal cyst · arachnoid cyst · skull fracture · hydrocephalus · posterior fossa

The term "leptomeningeal cyst" was coined in 1937 by Dyke¹ and was popularized in 1953 by Taveras and Ransohoff² to describe a kind of arachnoid cyst. Although it has been recognized that intracranial arachnoid cysts can be formed congenitally, or following trauma or inflammatory conditions,³,⁴,⁵,⁶,⁷,⁸,⁹,¹⁰ all leptomeningeal cysts have been thought to follow trauma. They usually occur in the parietal region associated with radiographic evidence of widening of a fracture and scalloping of the border of the inner table of the skull. At operation the dura over the center of the skull defect has invariably been found absent, and a cyst covered by arachnoid and filled with cerebrospinal fluid protruded through the bone defect.

Arachnoid cysts have been described as occurring in the posterior fossa, but leptomeningeal cysts in the posterior fossa are rare.

Case Report

An 18-year-old male student was admitted to Barnes Hospital on April 19, 1968, with complaints of occipital headaches and of a head tremor, both of which had been gradually increasing in severity for 5 years. His birth and development had been normal. At 2 years of age he had been struck by an automobile and had suffered a suboccipital midline skull fracture that extended to the foramen magnum, which rendered him unconscious for 3 days. He eventually recovered, and no other details of that illness are known. He had always seemed clumsy when playing.

Examination. The patient was alert and oriented and in no distress. There was a rhythmic vertical tremor of the head when he was erect. Bilateral horizontal nystagmus was evoked by lateral gaze, and there was mild papilledema. The right extremities were mildly dysmetric, and his gait was wide-based and ataxic. Tendon reflexes were normal, and Babinski responses were flexor. The routine blood studies and serum electrolytes were normal. Cerebrospinal fluid removed at the time of pneumoencephalography was also normal.

Radiological Studies. Plain films of the
skull demonstrated a large cystic lesion of the occipital bone which was posterior to the foramen magnum (Fig. 1). The lesion, which had sclerotic margins, appeared to arise in the diploë and expand inwardly. The inner wall was a thin shell. Tomographic examination of the occipital bone, using hypocycloidal tube movement, showed the cystic lesion to involve the inner table in an irregular fashion (Fig. 2). The lesion tapered at its margins to join the normal portions of the occipital bone. Pneumoencephalography (Fig. 3) showed a large cystic cavity that communicated with the subarachnoid space of the cisterna magna with free movement of cerebrospinal fluid and air in and out of the cavity. There was moderate dilatation of the entire ventricular system. The fourth ventricle was displaced forward. No air entered the prepontine space or passed over the cerebral convexities, indicating basilar arachnoiditis. A left brachial angiogram showed anterior displacement of the hemispheric branches of the posterior inferior cerebellar

Fig. 1. Plain films of the skull. Left: A cystic defect of the skull is seen extending from the posterior margin of the foramen magnum to the region of the internal occipital protuberance. Right: In the frontal projection, this defect has scalloped sclerotic margins and extends almost across the entire width of the occipital bone.

Fig. 2. Polytomographic examination of the occipital bone in the lateral (left) and axial (right) projections show the calcified inner margin of the cystic lesion. Noncalcified areas (asterisks) are present. Note the tapering at the margins of the lesion.

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Fig. 3. Pneumoencephalography shows free communication of the cystic lesion with the subarachnoid space. Note the spiculated appearance (open arrows) of the inner margin of the occipital bone. There is some anterior displacement of the fourth ventricle and moderate ventricular dilatation.

arteries. The vein of Galen was questionably elevated. There was no abnormal vascularity associated with the cyst. A right internal jugular venogram (Fig. 4) demonstrated anterior displacement of the confluence of the sinuses by the cystic lesion. There was no evidence of occlusion of the dural sinuses.

Intrathecal injection of radioactive iodine-tagged serum albumin showed the isotope to enter the cystic cavity and the lateral ventricles (Fig. 5). There was no accumulation of the iodinated albumin along the superior sagittal sinus at the end of 24 hours, which substantiated the diagnosis of basal arachnoiditis with an incisural block.

In summary, these radiological procedures indicated that there was a large epidural cyst which displaced the contents of the posterior fossa anteriorly, and which communicated freely with the subarachnoid space. There was a basilar arachnoiditis and an incisural block producing moderate hydrocephalus.

Operation. A posterior midline incision was used to expose the suboccipital region. Immediately after perforating the occipital bone, cerebrospinal fluid was encountered rather than dura. Removal of the remaining bone unroofed the cavity that had been seen radiographically. It was about 3 cm deep and was surrounded on all sides by bone. The in-

Fig. 4. Jugular venography shows anterior displacement of the confluence of the dural sinuses (arrow) by the cystic lesion.

Fig. 5. Intrathecal injection of radioactive iodine-tagged serum albumin. A scan at 24 hours shows accumulation of the RISA in the cystic cavity (open arrow) and in the ventricles (closed arrow). No activity is seen along the course of the superior sagittal sinus.
Fig. 6. Left: Operative field after removal of the occipital bone. In the center is the hole in the dura through which can be seen the foramen of Magendie (thin arrow). Broad arrow indicates calcified spicule on inner table of the intrasosseous cyst. Star indicates posterior arch of C-1. Right: The inner table of calcium has been removed and the dura (circles) opened. Broad arrow shows dual arachnoidal adhesions. Triangle indicates left cerebellar hemisphere. White arrow points to foramen of Magendie. Arrow with circle points to posterior inferior cerebellar artery.

The inner table of this cavity was about 1 mm thick and was adjacent to the anteriorly displaced dura. In the center of the inner table was a hole, 2 × 3 cm, through which the foramen of Magendie could be seen (Fig. 6 left). The inner and outer tables of this cavity were connected by calcified spicules. Other calcified spicules projected from both surfaces toward the center of the cavity, like stalactites on the walls of a cave (Figs. 3 and 6 left). The inner table of calcium was removed and the dura opened (Fig. 6 right), revealing a severe, generalized adhesive arachnoiditis. There were numerous dural-arachnoidal adhesions, and the foramen of Magendie was preserved as a hole within a sheet of fibrous tissue. The numerous, dense adhesions prevented further exploration. A silastic dural graft was used to effect a watertight closure. The suboccipital wound was closed and a low pressure ventriculoatrial Holter valve inserted.

Postoperative Course. The patient improved; however, the proximal end of the shunt had to be revised twice because of the formation of protein clots. He was last seen September 15, 1969. He no longer had headaches. His gait was still slightly ataxic, but was improving. He had returned to high school and was also working operating an offset press.

Discussion

We believe that the patient’s severe head injury in infancy caused both the epidural cyst and the chronic arachnoiditis, and that both of these were responsible for his symptoms.

The epidural cystic collection of cerebrospinal fluid, which eroded the occipital bone and which communicated freely with the fourth ventricle, is an unusual finding. In the only similar case reported, a multiloculated epidural cyst, which connected with the left lateral ventricle, was found in the left parieto-occipital-suboccipital area of a man who had sustained a head injury 50 years earlier; the interior wall of the cyst was lined by the inner table of the parietal and occipital bones and it extended below the tentorium, pushing both cerebellar hemispheres anteriorly.

We believe that the extradural lesion in our patient was a leptomeningeal cyst, which had been preceded by a skull fracture associated with a dural tear. We believe that at the time of the accident the occipital bone was fractured and the dura torn, the arachnoid of
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the cisterna magna then herniated through the dural tear and the fractured inner table of the thick occipital bone.

When this series of events occurs in the region of the thin parietal bone, which is a more characteristic location for leptomeningeal cysts, the whole bone is eroded and the margins of the skull defect are separated by a protrusion of an arachnoid-lined cyst over which there is no dura.²,⁶,¹⁰,¹³,¹⁵,¹⁸,²⁰,²⁴

Taveras and Ransohoff²¹ postulated that in these cases the dura is torn at the time of fracture and the pulsating arachnoid gradually herniates through the dura and erodes the bone.

In our patient the fracture involved the much thicker occipital bone. The arachnoid did herniate through the fracture, but not through the entire thickness of the bone, only through the inner table. With time, the increased intracranial pressure produced by the trauma, arachnoiditis, and constant pulsations of the arachnoid resulted in an intrasosseous enlargement of the suboccipital area. This proposed mechanism would explain the presence of a posterior fossa extradural cyst which caused radiographic ballooning and scalloping of the suboccipital bone and which communicated freely with an enlarged fourth ventricle.

Other explanations for this lesion are possible, but we believe they are less tenable. Arachnoid cysts may act as mass lesions anywhere within the cranium, but other than the leptomeningeal variety, they occur subdurally.⁵,¹⁰,¹²,¹³,¹⁶,¹⁸,¹⁹,²⁰,²²-²⁵,²⁷

Another possibility might be that this started as an epidural hematoma, but we believe such an explanation unlikely. Our patient did have an occipital skull fracture at the time of his initial injury, and epidural hematomas are commonly associated with such fractures.¹²,¹⁷,²⁹ In this case, a chronic epidural hematoma would have to have become calcified to account for the calcified membrane outside the dura, and with time there would have been ballooning of overlying bone, which can occur with any slow-growing intracranial lesion.²⁰ However, those chronic epidural hematomas that have been reported¹⁵,⁷,¹₄,²₂,²⁶,²⁸ have usually produced symptoms for days or weeks, but rarely years. We have found only three cases of epidural hematomas which became calcified and all of these were supratentorial. One was presented as a radiologic abnormality²² and no clinical history was given. A second one existed 15 months,²⁸ and the third may have existed 6 years.⁷ Also, these calcified hematomas²,²⁵ contained unmistakable degradation products of blood and fibrin clot, and resembled calcified subdural hematomas,¹⁰ which was not the case in our patient. The two most chronic cases of epidural hematoma of the posterior fossa reported probably existed 14 years¹ and 9 months,²³ and neither of these calcified. Ventriculograms in both these patients showed that neither communicated with the ventricles and both showed a lack of filling of the fourth ventricle, which one might expect with an extraxial mass that does not communicate with the ventricle. In our patient, on the contrary, there was not only communication with the fourth ventricle, but also the fourth ventricle was enlarged.

References

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