Epidermoid cyst in the brain stem

Case report

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A patient is reported with an epidermoid cyst of the brain stem, presenting as a progressive brain-stem mass, together with recurrent aseptic meningitis.

Key Words • epidermoid cyst • brain stem

Intracranial epidermoid cysts account for only 1% of all intracranial tumors. The great majority of cases are associated with congenital dermoid sinuses and recurrent episodes of septic meningitis. We describe an apparently unique case of an epidermoid cyst occurring in the brain stem.

Case Report

This 3½-year-old girl was referred to us in October, 1975, with increasing drowsiness and bilateral papilledema, following a third episode of meningitis.

History. At the age of 14 months, she had been admitted to another hospital with a 2-day history of cough and fever associated with vomiting. She was irritable and had meningism. The cranial nerves were intact but bilateral Babinski signs were present. Lumbar puncture revealed clear cerebrospinal fluid (CSF) under normal pressure. Laboratory analysis, including cell and protein studies, was normal and no organisms were cultured. Antibiotic therapy was begun, but 3 days later she was noted to have developed a dense left hemiparesis, not involving the face. Again lumbar puncture was normal. Blood cultures were negative. She gradually improved and was discharged home 3 weeks later with a mild left hemiparesis.

One year later, the patient was readmitted with malaise, vomiting, and drowsiness, and on examination again had meningism. There was still evidence of a mild left hemiparesis, but on this occasion a definite right, lower motor neuron facial paresis was also present. Lumbar puncture revealed cloudy CSF, with 180 white blood cells (WBC's), mainly polymorphonuclear leucocytes, 60 mg% protein, and 35 mg% glucose. No organisms were recovered. She was again treated with antibiotics, and was able to return home 3 weeks later, still with evidence of right facial weakness and left hemiparesis. Soon after discharge she developed a squint, apparently due to a sixth nerve palsy.

The patient was readmitted 6 months later with an identical presentation. The weaknesses previously noted were still present. Lumbar puncture revealed cloudy CSF, with 3395 WBC's, mainly polymorphonuclear leucocytes, 135 mg% protein, and 50 mg% glucose. Again no organisms could be recovered.

Examination. She looked ill and was stuporous, but responded to simple verbal commands. Bilateral papilledema was noted,
FIG. 1. Dimer-X ventriculography, lateral view, showing defect in the floor of the fourth ventricle.

together with a mild right sixth nerve paresis. Bilateral facial pareses were noted, worse on the right; mild left hemiparesis with bilateral extensor plantar reactions persisted. Dermal sinuses and stigmata of congenital disease were specifically looked for but not found. Ventriculography showed an apparently lobulated filling defect in the floor of the fourth ventricle (Fig. 1).

First Operation. A posterior fossa exploration was carried out. The cerebellar tonsils were separated and the floor of the fourth ventricle was found bulging tensely backward; it was pale in appearance and felt cystic. A small area of the ventricle floor was coagulated with bipolar diathermy, and, as there were no changes in the vital functions, a blunt cannula was inserted. At a depth of 2 mm a cystic cavity was entered and 20 ml of creamy fluid was aspirated. The floor of the fourth ventricle collapsed forward. The cavity was irrigated with antibiotic and finally 0.5 ml of micropaque barium sulphate (Steripaque) was instilled. The rest of the cavity of the fourth ventricle was protected with patties. A small biopsy was taken from the wall of the cyst, but this revealed no specific tissue. No organisms were recovered from the fluid. Abundant epithelial cells were noted.

First Postoperative Course. The patient's condition was immediately improved. She became fully conscious and obeyed all commands. The following day she was able to swallow satisfactorily. The pupils were normal but the eyes were deviated toward the left. The facial pareses and the left hemiparesis remained unchanged. There was no evidence of temperature or meningism.

A postoperative skull film showed the cavity in the brain stem outlined by Steripaque, just to the left of the midline and measuring 3 × 2 × 2 cm. She was discharged home 3 weeks later, with the same pattern of neurological deficit, although it was rapidly reducing.

Three months later the patient was admitted to another hospital with pneumonia, clearly due to progressive bulbar palsy and later transferred to us in a state of deepening coma. Plain x-ray films showed that the barium-outlined cavity had greatly increased in size. Exploration revealed the same features as previously.

Second Operation. Radical excision was performed and the epidermoid cyst was found to extend completely through the brain stem to the prepontine subarachnoid space. A total removal was considered to have been achieved. Histology of the excised cyst showed a squamous cell-lined cyst wall with no ancillary skin structures, that is, an epidermoid cyst.

Second Postoperative Course. The postoperative course was extremely stormy with an initial improvement in consciousness, but continuing bulbar problems, later accompanied by CSF leakage from the wound and infection. She died 2 months after the second operation.

Postmortem Examination. Autopsy confirmed the operative findings, namely that the brain stem was completely split (Fig. 2). No
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epidermoid cyst remained. The basilar artery together with the prepontine arachnoid was applied to the pons as a membrane, suggesting that the epidermoid cyst arose behind the artery. Death appeared to have been due to a posterior fossa abscess.

**Discussion**

Epidermoid and dermoid cysts are usually considered together since their natural histories are so similar and their separation often depends only on the microscopic appearance of the cyst wall.6,8,9

Dermoid cysts are more common in the midline and epidermoid cysts are most often found in the lateral aspect of the intracranial cavity.5,9 but they have been found in the cerebellopontine angle, parapituitary region, diploë, rhomboid fossa, intraspinally, intracerebrally, within the cerebellum, and in the ventricular cavities.7-9 The epidermoid cyst in this instance appeared to arise within the brain stem with a thin layer of ependyma posteriorly and a thin arachnoid membrane anteriorly, although it is possible that it could have arisen in the prepontine subarachnoid space and extended back into the brain stem.

The presentation of the tumor can be as a space-occupying lesion or as a source of recurrent meningitis.2,8,5 The meningitis can be bacterial, in which case a congenital dermoid sinus is usually present,1 or aseptic, as in this case.

The treatment of choice is surgical excision. Special care should be taken on opening the cyst in order to avoid spilling the contents into the subarachnoid space, for a violent reactive meningitis can occur under such circumstances.6 Matson4 recommended the total excision of the dermal tract and the cystic extensions wherever they occurred. Some tumors, such as that in our case, may pose considerable surgical problems because of their treacherous anatomical location, and as a result subtotal removal is often all that is achieved. The risks of such palliation have been clearly illustrated in our case. We now regret not having attempted radical resection at the initial operation. There was at that time, however, a slight diagnostic equivocation regarding the possibility of the cyst being an abscess.

Our case has special clinical interest because, as a result of the anatomical site of the cyst, presentation was that of a progressive space-occupying lesion and involved recurrent episodes of aseptic meningitis.

**References**


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