Epidermoid cysts of the posterior fossa

MITCHEL S. BERGER, M.D., AND CHARLES B. WILSON, M.D.

Department of Neurological Surgery, School of Medicine, University of California, San Francisco, California

Epidermoid cysts originating in the paramedian basal cisterns of the posterior fossa are congenital lesions that grow to a large size through slow accumulation of desquamated epithelium. These lesions grow between and ultimately displace cranial nerves, vascular structures, and the brain stem, causing a long course of progressive neurological deficits. The onset of symptoms usually occurs during the fourth decade of life. Epidermoid cysts are easily diagnosed with computerized tomography scans, which characteristically show a low-density extra-axial pattern. The primary surgical objective is to decompress the mass by evacuating the cyst contents and removing nonadherent portions of the tumor capsule; portions of the capsule adherent to vital structures should be left undisturbed. Aseptic meningitis is the most common cause of postoperative morbidity, and its incidence may be minimized by intraoperative irrigation with steroids followed by systemic therapy with dexamethasone. Symptomatic recurrences that occur many years after surgery should be managed with conservative reoperation.

KEY WORDS • epidermoid cyst • aseptic meningitis • computerized tomography • posterior fossa

Epidermoid cysts that originate in the lateral subarachnoid cisterns of the posterior fossa are slow-growing extra-axial lesions that gradually produce symptoms and signs of an expanding mass. Characteristically, epidermoid cysts spread across the basal surface of the brain, traveling along natural planes, and in the later stages wrap around and adhere to critical neurovascular structures. Although computerized tomography (CT) has facilitated the diagnosis of epidermoid cysts, the full extent of involvement often cannot be determined until surgery. In this report, we present a series of patients with epidermoid cysts treated with conservative, rather than radical, surgery.

Summary of Cases

Patient Population

Between 1972 and 1983, 13 patients with epidermoid cysts of the paramedian posterior fossa underwent surgery at the University of California, San Francisco. There were seven men and six women, aged 21 to 74 years (mean 47 years). The onset of symptoms occurred 3 months to 20 years before the operative procedure (mean 4.3 years). Three patients complained of trigeminal neuralgia involving the lower half of the face, but no other signs of cranial nerve compression were present. One patient had typical hemifacial spasm without involvement of the trigeminal nerve roots. The remaining patients had symptoms and signs of an expanding mass in the cerebellopontine angle that extended through the incisura, including diplopia, nystagmus, vertigo, decreased hearing, facial weakness, hemiparesis, disturbance of gait, and appendicular ataxia. One patient had two bouts of aseptic meningitis before radiographic confirmation of an epidermoid cyst was obtained (Table 1).

Radiographic Studies

All but one of the patients underwent CT evaluation with contrast enhancement. Three patients also received 5 ml (170 mg/ml) of intrathecal metrizamide to further define the extent of the tumor. In the remaining patient, who was evaluated before the advent of CT, skull radiography, planar tomography, and pneumoencephalography were used in the diagnostic evaluation.

On CT scans, the lesions were consistently hypodense, with attenuation values slightly above zero, and often extended into the middle fossa (Figs. 1 and 2). One epidermoid cyst had a small area of higher density (30 Hounsfield units) and calcifications were present in two, but none of the lesions showed contrast enhance-
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Fig. 1. Axial computerized tomography scan obtained after intravenous infusion of contrast material. The parapontine cisterns are asymmetrical as a result of a low-density nonenhancing mass on the left (large arrows). The epidermoid cyst has eroded the petrous apex on that side and extends into the medial aspect of the left middle fossa (small arrows).

Fig. 2. Axial computerized tomography scan through the tentorial incisura showing contiguous spread of the epidermoid cyst from the posterior fossa to the hippocampal region (arrows).

ment. In one patient, metrizamide was particularly helpful in determining the superior, inferior, medial, and lateral extent of the tumor (Fig. 3). In the patient who had recurrent aseptic meningitis, CT scans revealed isolated hypodense deposits over the convexity and in the ambient cisterns; these deposits were considered to be portions of the ruptured cyst (Fig. 4). The liquid cystic center, which is present in a minority of epidermoid tumors, occupied a small part of this lesion.

Operative Technique

The lesion was approached through a suboccipital craniectomy in 12 patients and through a frontotemporal craniotomy in one. In one patient, the initial subtentorial approach failed to provide adequate exposure, so a second procedure was performed within 30 days through a frontotemporal craniotomy. In all cases, the glistening tumor capsule adhered to portions of the brain stem and subarachnoid neurovascular structures, and finger-like protrusions of the epidermoid cyst insinuated themselves between the cranial nerves and the basilar cisterns. After the capsule was incised, the flaky white contents were removed with meticulous care and nonadherent portions of the capsule were excised. Complete resection of the adherent capsule was not attempted in any case. In one patient who had an extradural tumor that extended into and eroded the petrous apex, a gross total resection was obtained. The cavernous medial temporal region was further exposed, if necessary, by opening the tentorium. After removal of the cyst contents, the adjacent cisterns were irrigated with large volumes of a hydrocortisone solution (100 mg/liter of lactated Ringer's solution) to remove any potentially irritating debris. In two of the three patients with trigeminal neuralgia and in one patient with hemifacial spasm, the epidermoid cyst had displaced an arterial loop into the root entry zone of the corresponding cranial nerve. In these patients, the vessel was dissected from the nerve after the tumor contents were evacuated.

<table>
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<tr>
<th>Lumbar Puncture</th>
<th>White Blood Cells (/cu mm)</th>
<th>Monocytes (%)</th>
<th>Lymphocytes (%)</th>
<th>Glucose (mg%)</th>
<th>Total Protein (mg/dl)</th>
<th>Opening Pressure (mm H2O)</th>
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<td>128</td>
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FIG. 3. Left: Axial computerized tomography scan obtained after intrathecal instillation of metrizamide. The basilar artery (large arrow) is displaced away from the lesion (small arrows), which is outlined by the metrizamide. Right: Coronal reformation through the lesion shown left. Metrizamide fills the interstices of the epidermoid cyst (crosses) and demarcates its superior (large arrow) and lateral (small arrow) borders.

FIG. 4. Axial computerized tomography scan showing a small low-density deposit of material (arrow) from the ruptured cyst over the right frontal convexity.

Postoperative Course

Postoperative morbidity from manipulation of cranial nerves was transient in all patients, except one with hemifacial spasm who developed a hearing loss. Two patients had pseudomeningoceles and communicating hydrocephalus that required shunting of cerebrospinal fluid (CSF). Another patient developed tension at the operative site, which resolved after 2 weeks of treatment with steroids, acetazolamide (Diamox), and compression dressings. This patient and one other patient had mild aseptic meningitis, which responded promptly to dexamethasone treatment. Purulence at the incision site was treated with local wound care in one patient and required debridement in another.

The mean follow-up period is 4½ years (range 9 months to 12 years). All 13 patients are clinically stable, and none has required a subsequent operation for regrowth of the epidermoid cyst. The patients with trigeminal neuralgia and hemifacial spasm have not had recurrence of their symptoms during a mean follow-up interval of 3½ years. The patient who developed aseptic meningitis before surgery has not had another episode.

Discussion

Intracranial epidermoid cysts arise from displaced epithelial tissue between the 3rd and 5th weeks of gestation during closure of the neural tube. Although separation of neuroectoderm from its cutaneous counterpart occurs dorsally along the midline, the lateral location of most epidermoid cysts may be explained by proliferation of multipotential embryonic cell rests. Alternatively, epithelial rests may be carried into the region of the cerebellopontine angle with the developing otic vesicles.

The lesion is composed of desquamated epidermal cell debris originating from an inner epithelial lining of the glistening white tumor capsule. The desquamated epithelial cells are rich in cholesterol and, although epidermoid cysts are usually solid, they may have a liquid cystic center. Dermoid cysts, which contain elements of dermal cell layers (namely, hair and sebaceous glands) not present in epidermoid tumors, have a different consistency and lack the milky-white appearance of epidermoid cysts. Dermoid cysts are less common lesions and tend to be located in the midline.

Intracranial epidermoid cysts constitute 0.2% to 1.8% of all intracranial tumors, and occur equally in males and females. The onset of symptoms...
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usually occurs in patients aged between the mid-20's and the early 50's. Unlike dermoid cysts, which are accompanied by other congenital anomalies in up to 50% of patients, epidermoid cysts are isolated lesions. They are found in several intracranial locations and, through contiguous spread along normal cleavage planes, may extend through the tentorial incisura and occupy more than one site. The area most often involved is the basal surface of the brain (subarachnoid cisterns), particularly the cerebellopontine angle and the parasellar region. Epidermoid cysts are also found in the fourth ventricle, cerebellum, cerebral hemispheres, brain stem, and lateral ventricles. In one of our patients, extradural extension into the trigeminal region caused erosion of the petrous apex; a similar case had been described by others.

Epidermoid cysts grow slowly through gradual accumulation of normally dividing cells and often attain a large size before the onset of symptoms. Unlike most neoplastic lesions, which have an exponential growth, epidermoid cysts grow at a linear rate that resembles the growth of human epidermis. Therefore, the duration of symptoms and signs is typically prolonged, ranging from several months to 30 years. In one report of this disease, 20% of patients had symptoms for 10 to 25 years. As more series are collected with CT data, the mean duration of symptoms should approach that in our patients (4.3 years).

Intracranial epidermoid tumors that are isolated to the posterior fossa or extend through the incisural cause symptoms by gradually compressing and eventually surrounding cranial nerves and the adjacent brain stem. These lesions often adhere to blood vessels, and for that reason were once classified by Lepoir and Pertuiset according to their relationship to the basilar, carotid, and choroidal arteries. Epidermoid cysts originating in the subarachnoid cistern of the cerebellopontine angle cause symptoms and signs of a slowly expanding mass in that region, including ataxia, nystagmus, facial hypesthesia, aresthesias and weakness, hypoacusis, and hemiparesis. Epidermoid cysts of the posterior fossa have been known to cause trigeminal neuralgia, atypical facial pain, glossopharyngeal neuralgia, and hemifacial spasm. Two of our four patients with trigeminal neuralgia and hemifacial spasm reported that tumors causing tic douloureux "do so by pushing the trigeminal nerve against the blood vessel." This mechanism was substantiated in three of our four patients with trigeminal neuralgia and hemifacial spasm. Other authors consider a direct compressive effect of the mass near the root entry zone to be the causative factor. Two of the larger reported series, however, were collected before the operating microscope was developed and the presence of an offending vascular loop might possibly have been overlooked.

Unlike patients with dermoid cysts, who may have repeated bouts of bacterial meningitis, patients with epidermoid cysts may have brief recurrent episodes of aseptic meningitis caused by rupture of the cyst contents. During the ictus, the patient has fever and meningeval irritation, accompanied by pleocytosis, decreased glucose and elevated protein levels, and negative CSF cultures. A variant of this entity is Mollaret's meningitis, or aseptic leukocytoid endothelial meningitis; although the symptoms and signs resemble those of aseptic meningitis, it is usually of unknown etiology and associated with the early appearance of large delicate endothelial cells in the CSF. We were unable to find these epithelial-like cells in our patient with recurrent aseptic meningitis; however, there have been reports of Mollaret's meningitis associated with an intracranial epidermoid cyst.

High-resolution CT with thin-section axial images through the posterior and middle fossa is the optimal diagnostic procedure for evaluating epidermoid cysts. Often, coronal reformations can further delineate the incisural and parasellar components of the lesion, and intrathecal injection of metrizamide can help to define the extent of tumor. Epidermoid cysts are low-density lesions that have attenuation values slightly greater than that of CSF and do not enhance with intravenous contrast agents. Some intracranial epidermoid cysts show increased density on CT scans that is consistent with saponification of the cystic contents or hemosiderin deposits. Epidermoid cysts rarely contain areas of calcification; when present, such areas are often located in the margins of the lesion. Regardless of the density of the lesion on CT scans or the presence of a calcified rim, contrast enhancement is distinctly abnormal and suggests a malignant epithelial component.

The surgical treatment for epidermoid cysts in the posterior fossa, including those that extend above the incisura, should be conservative. Unattached portions of the tumor capsule should be excised and the soft flaky contents removed; a dental mirror may be used as required for visualization. Although this approach does not result in a surgical cure, it will alleviate the patient's symptoms and prevent progression of the disease for many years. Because the capsule is nearly always adherent to vital neurovascular structures, complete removal is unwise and should be avoided. Most epidermoid cysts are readily accessible through a suboccipital craniectomy; if necessary, further exposure of the parasellar region can be gained by cutting the incisural margin. The surgical approach depends upon the extent of the tumor as determined by the preoperative diagnostic studies. If a significant portion of the lesion is located in the parasellar medial temporal region, a subtemporal-transiental route should be chosen.

Postoperative aseptic meningitis due to leakage of irritating debris into the basal cisterns may occur despite complete removal of the cyst contents and much of the capsule. Steroids promptly alleviate these symptoms and antibiotics should be administered until the CSF culture is verified as negative. We add hydrocortisone to the irrigating solution during the
operation and institute systemic steroid therapy afterward, gradually tapering the dexamethasone dose over 3 weeks after the patient is discharged from the hospital. Two of our patients subsequently developed communicating hydrocephalus, which has been reported to be a sequel of aseptic (chemical) meningitis.\(^2\)\(^{,}\)\(^{23}\)

Despite their slow growth, epidermoid cysts can recur, presumably from the residual tumor capsule.\(^5\)\(^{,}\)\(^{17}\)\(^{,}\)\(^{20}\)\(^{,}\)\(^{48}\)

The potential for regrowth of a congenital tumor must be recognized over a period of time equivalent to the patient’s age at onset of symptoms plus 9 months.\(^2\) As most patients become symptomatic during their fourth decade of life, it may take 30 to 40 years for recurrent symptoms to develop, underscoring the need to avoid radical excision. However, if the lesion recurs soon after partial resection and displays contrast enhancement on the CT scan, malignant degeneration or infection may be present, necessitating further resection or debridement, respectively.\(^8\)\(^{,}\)\(^{34}\)\(^{,}\)\(^{39}\) Postoperative radiation therapy for benign epidermoid cysts is not indicated, as recurrence has been documented with its use.\(^2\)\(^8\)

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Address reprint requests to: Mitchel S. Berger, M.D., c/o The Editorial Office, 1360 Ninth Avenue, Suite 210, San Francisco, California 94122.