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Pineal epidermoid cysts: diagnosis and management

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Object. The results of surgical treatment of epidermoid cysts of the pineal region in six cases are presented.

Methods. Six patients with pineal epidermoid cysts underwent surgery at the Institute of Neurosurgery "N. N. Burdenko," in Moscow, during the period 1976 to 1995. The duration of the patients' preadmission clinical history varied from 6 months to 2 years (average 1.4 years). Headache, diplopia, and vertigo were the most frequently occurring symptoms. Neurological examination demonstrated papilledema, impaired pupillary reaction, ataxia, and long-pathways deficit; Parinaud's syndrome was found in only one case. Computerized tomography and magnetic resonance imaging constituted the primary diagnostic tools. Surgery was performed using either an infratentorial–supracerebellar approach (two cases) or an occipital–transtentorial approach (four cases).

Conclusions. Surgical results can be excellent if a removal, as extensive as possible, is performed using either the supracerebellar or occipital–transtentorial approach.

KEY WORDS • **epidermoid cyst** • **pineal region** • **computerized tomography scanning** • **magnetic resonance imaging** • **supracerebellar approach** • **occipital–transtentorial approach**

EPIDERMOID cysts are relatively rare, congenital lesions that account for approximately 1% of all intracranial space-occupying lesions.[8–10,19,20,22,23,31,32] Together with intracranial dermoid cysts, they are thought to originate from the germinal layer and ectopic nests of epithelial cells left in situ during closure of the neural tube.[24,30] Epidermoid and dermoid cysts may be clearly distinguished from one another by their location; the latter show a midline localization whereas the former are usually located eccentrically.[9,23,35]

Epidermoid cysts of the pineal region are very rare.[12,15] In an extensive review of the literature on intracranial epidermoids, Netsky[17] reported on only two cases. Two possible additional cases were noted by Yaşargil, et al.,[35] in their extensive study of epidermoid and dermoid cysts, and other sporadic cases have been noted by other authors.[15,23,26,27,34] In the last 9 years at the Institute of Neurosurgery, N. N. Burdenko, we have performed surgery in six patients in whom epidermoid cysts were located primarily in the pineal region.

Clinical Material and Methods

The present six cases account for 3.1% of the 133 intracranial epidermoids and 3.4% of the 175 pineal tumors surgically treated at our institution from 1976 through 1995. Patient genders were equally represented—three males and three females—and the average age was 30 years (range 19–40 years; Table 1).

The duration of the patients' preadmission clinical history varied from 6 months to 2 years and averaged 1.4 years. Headache was a constant complaint in this series of patients and, as a rule, was described as dull and increasingly severe. Diplopia was a complaint in three patients and vertigo and/or gait imbalance was a complaint in three patients. Interestingly, two of the latter three patients also reported experiencing hearing impairment, which subsided following the operation (Table 2).

The results of the neurological examinations performed at admission are summarized in Table 2. Papilledema was observed in most cases, whereas Parinaud's syndrome—a typical sign of pineal tumors—was detected in only one patient.

Computerized tomography (CT) and magnetic resonance (MR) imaging represented the chief diagnostic tools. Angiography was not performed in any of the present cases. Surgery was performed using either an infratentorial–supracerebellar approach (Cases 1 and 3) or an occipital–transtentorial approach (Cases 2, 4, 5, and 6). Because there was diagnostic suspicion of a brainstem tumor, direct surgery was preceded by stereotactic biopsy in one case (Case 5; Fig. 1).

Results

Computerized Tomography Scanning Data

As a rule, the lesion appeared as a round or oval homogeneous low-density mass that did not enhance after administration of a contrast agent. The mass generally displayed well-defined margins, no calcification, and no perifocal edema (Fig. 2). A certain degree of ventricular en-

TABLE 1

Characteristics in six patients with pineal epidermoid cysts

Case No.	Age (yrs), Sex	Follow Up (mos)
1	19, F	72
2	39, F	52
3	32, M	53
4	40, F	64
5	19, M	59
6	24, M	37

largement was present in all cases. Periventricular lucency, a sign of cerebrospinal fluid (CSF) resorption caused by obstructive hydrocephalus, was detected in two cases.

Magnetic Resonance Imaging Results

In all but one case MR imaging revealed a mass with well-defined margins that was nonhomogeneous and hypointense on T_1-weighted images and exhibited a definite increase in proton density on T_2-weighted spin-echo images. In the remaining patient (Case 6), the lesion appeared to be almost isointense on T_1-weighted images, although it still appeared hyperintense on T_2-weighted spin-echo images (Fig. 3).

Surgical Findings

A supracerebellar approach, which was used in two patients (Cases 1 and 3) treated early in the series, allowed convenient exposure of the lesion and radical removal of the lesion in one of the two cases.

Another patient (Case 2) treated early in the series underwent surgery in the "three-quarters prone position" via an occipital–transtentorial approach described by Ausman, et al.[2] During the operation, the unfamiliar view of the surgical anatomy and our inability to visualize contralateral basal structures rendered this operation rather difficult. We had to leave in situ a tiny fragment of the capsule that was adherent to the vein of Galen. We used a mirror extensively to check contralateral blind corners so that we could prevent tumor fragments from being left behind.

The remaining three cases were surgically treated via an occipital–transtentorial approach performed with the patient in the sitting position. In Cases 4 and 5 definite tumor extension toward the infundibular recess of the third ventricle was observed and, thus, these patients were judged unfit to undergo an infratentorial approach. In Case 6 there was significant supratentorial involvement, and the cyst pushed down the entire deep venous system, including the vein of Galen itself, which made us choose an approach from above (Fig. 4). Except for Case 2, convenient exposure of the lesion and important surrounding structures was obtained in all cases. Small fragments of the capsule had to be left in situ in three cases (50%) because of their tenacious adherence to the deep veins (Fig. 5). A radical removal was achieved in the remaining cases.

Clinical Results

In one case, a 2-week course of steroid medication was required for postoperative aseptic meningitis to subside. No other complications were recorded.

TABLE 2

Symptoms and neurological signs in six patients with pineal epidermoid cysts

Symptom/Sign	No. of Cases (%)
symptom	
headache	6 (100)
diplopia	3 (50)
vertigo &/or dizziness	3 (50)
decreased hearing	2 (33)
neurological sign	
papilledema	5 (83)
impaired pupillary reaction	3 (50)
ataxia	3 (50)
long-pathways deficit*	3 (50)
nystagmus	2 (33)
Parinaud's syndrome	1 (17)
parkinsonism†	1 (17)

* Hemisensory deficit.
† Bilateral akinesia and rigidity.

Postoperative CT scanning was performed in all cases at an average of 6 months postsurgery and revealed some residual fragment of the capsule in only one case. Computerized tomography cisternography was also performed in one case to rule out a recurrent tumor that had not been fully excluded by control plain CT scanning (Fig. 6). All patients are presently asymptomatic 3 to 6 years (average 56 months) following surgery.

Discussion

Intracranial Epidermoid Cysts

Intracranial epidermoid cysts account for approximately 1.5% (range 0.5–1.8%)[3,9,19,32] of all intracranial masses described in different series. "Dumbbell" supra/infratentorial lesions occur infrequently but are not exceptional,[9,23,35] as also are those located in the fourth ventricle.[3,7,16,23,26] The pineal region is an extremely rare site for intracranial epidermoid cysts to occur.[12,15,17,27] In two recent large studies investigators reported two cases each of pineal epidermoid cysts in approximately 40 cases of intracranial epi-

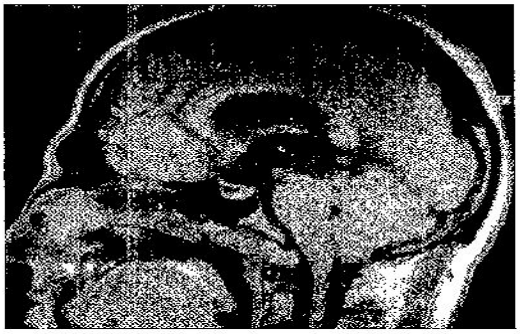

FIG. 1. Case 5. Sagittal T_1-weighted MR image demonstrating a round, hypointense lesion located in the posterior portion of the third ventricle, which does not show clear demarcation from the quadrigeminal plate.

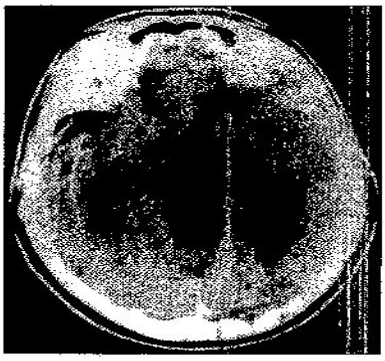

FIG. 2. Case 2. Computerized tomography scan demonstrating a round, hypodense mass in the pineal region displacing the posterior portion of the third ventricle.

dermoids in each series.[23,35] This matches well with our observed rate of occurrence of pineal epidermoid cysts in all intracranial epidermoids.

Pineal Epidermoid Cysts

In our series, as well as in reported cases in the literature, the absence of gender prevalence and the relatively young age of the patients (average 30 years) matches that of intracranial epidermoid cysts as a whole.[1,9,23,26,35] However, the duration of clinical history appears to be shorter than that of epidermoids in other locations: 1.4 years on average in the present series. This is explained by the tendency to cause obstructive hydrocephalus, which, in general, is shown by epidermoids of the pineal region.[34] In fact, symptoms and signs of increased intracranial pressure caused by hydrocephalus represent the chief clinical characteristic of pineal epidermoids. Upward-gaze paresis, a typical sign of pineal tumors,[18] is rarely detected in pineal epidermoid cysts; it was found in one of the six present cases. However, more subtle symptoms of local mass effect such as pupillary abnormalities were detected

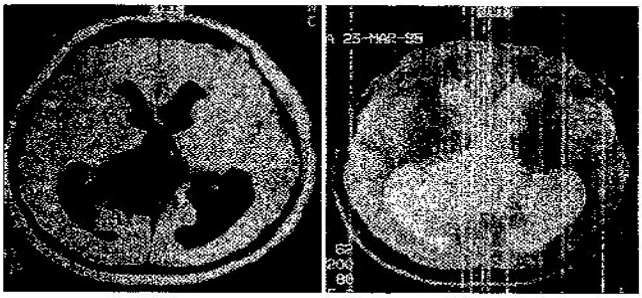

FIG. 3. Case 6. Magnetic resonance images. *Left:* A T_1-weighted image revealing a huge, slightly hypointense dishomogeneous lesion displacing and occupying the posterior portion of the supratentorial ventricular system. *Right:* A T_2-weighted image displaying a homogeneous high signal.

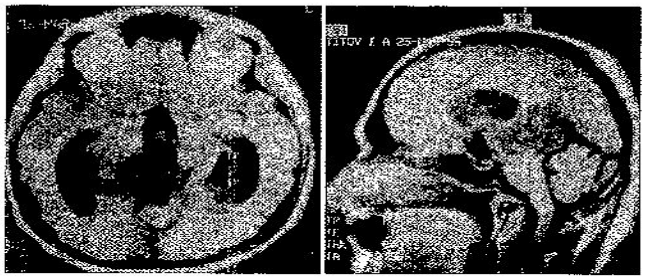

FIG. 4. Case 6. Preoperative T_1-weighted MR images demonstrating inferoposterior displacement of the deep venous system.

in a higher percentage of cases, as were symptoms of cerebellar dysfunction.

Interestingly, in two of the present cases, the patients complained of decreased hearing that definitely improved after surgery. We had already observed this unusual (for pineal masses) sign in another subgroup of benign pineal masses, meningiomas.[13] DeMonte and colleagues[5] have extensively discussed the pathophysiological mechanism underlying hearing impairment in the presence of pineal tumors.

Diagnostic Neuroradiology of Pineal Epidermoid Tumors

Modern radiological diagnosis of pineal tumors is based on findings on CT and MR imaging. Angiography is no longer recommended routinely in these cases, although its value in delineating the anatomy of displaced deep venous system structures and tumor vascularization is unquestionable.[15] As a rule, CT scanning demonstrates a mass that occupies the quadrigeminal cistern, causes hydrocephalus, and exhibits low density, a round or oval shape, no contrast enhancement, and the absence of perifocal edema.[6,11,17,21,26] Magnetic resonance imaging can offer some additional clues for the proper preoperative diagnosis and is superior to CT scanning in delineating the actual relationships between the mass and surrounding neurovascular structures. Epidermoids of the pineal region appear as cauliflower-shaped masses that compress the posterior third ventricle and the sylvian aqueduct and do not enhance after administration of a paramagnetic contrast agent. They are typically hypointense on T_1-weighted images, whereas their proton density definitely increases on the T_2-weighted spin-echo images, and they do not enhance in response to administration of a paramagnetic contrast agent.[4,14,25,28,33,35] With this diagnostic modality, CSF-containing cavities in the site of previously removed epidermoid cysts can be distinguished from recurrent epidermoids,[25] thus obviating the need to perform CT cisternography. However, we had to perform CT metrizamide cisternography in one of the present cases to rule out definitely a recurrent tumor. Olson and associates[20] outlined the possibility that MR imaging is not capable of providing a preoperative diagnosis in intracranial epidermoids. Those located in the pineal region can be confused with intraaxial tumors and their intraarachnoid invaginations and irregular margins can sometimes be wrongly inter-

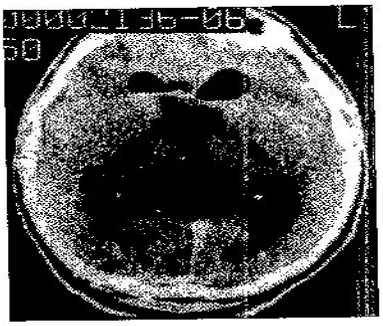

FIG. 5. Case 6. Postoperative CT scan revealing a very thin fragment of capsula as a small remnant of the surgically treated lesion.

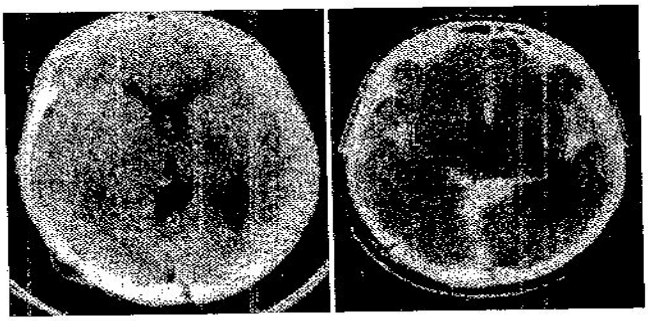

FIG. 6. Case 2. *Left:* Postoperative CT scan obtained at the level of the quadrigeminal plate revealing an eccentric calcification located at the periphery of a round cavity. It is unclear whether this represents a tumor remnant. *Right:* Computerized tomography cisternography demonstrating filling of the cavity by free-flowing CSF.

preted as indicating an infiltrative character. Actually, in one of the present cases, stereotactic biopsy was performed because there was suspicion of intraaxial tumor. We did not perform MR angiography in any of the present patients. However, that examination appears to be a promising tool for cases of pineal tumors in general, and epidermoids in particular, because it may clearly delineate the dislocated deep venous system, thus providing important information for surgical planning.

Surgical Management of Pineal Epidermoids

In our cases we used the most commonly performed approaches for surgical management of pineal tumors in the microsurgical era.[2,13,29]

In a previous publication[13] we mentioned our difficulties in using the three-quarter prone position,[2] which we used in one patient in the present series. In that case, intraoperatively we extensively used a dental mirror so that we could examine blind corners. We had the impression that we had achieved total removal of the lesion, although we could not be absolutely certain of this. Postoperatively, we had to perform CT cisternography to exclude the presence of residual tumor.

In the remaining cases, we used both the infratentorial–supracerebellar and the occipital–transtentorial approaches. In the two cases in which we used the infratentorial–supracerebellar approach, the tumor was seen well before the veins came into view, and removal of the lesion proceeded uneventfully. We used the occipital–transtentorial approach in the last three cases, in which supratentorial extension of the lesion would not have allowed us to obtain an adequate view of the mass if approached from below the tentorium. Radical removal of the lesion including the capsule, which was possible in 50% of the present cases, was unrelated to the approach used but was achieved when significant adhesions between the capsule and the veins of the galenic system were not encountered. In cases in which significant adhesions were present, we deliberately left in situ fragments of the capsule intimately adherent to the deep veins to prevent any undue risk of injuring them.

Our experience would indicate that both the infratentorial–supracerebellar and the occipital–transtentorial approaches can be used in the treatment of pineal epidermoids. We would suggest reserving the supratentorial approach for those lesions having a significant supratentorial component. Yaşargil, et al.,[35] have mentioned both these approaches as being indicated in the surgical management of pineal epidermoids.

Conclusions

Pineal epidermoid tumors are rare, slow-growing, extraaxial benign lesions that exhibit a shorter duration of clinical history than intracranial epidermoids in other locations because of their tendency to compress the sylvian aqueduct and cause obstructive hydrocephalus. Clinical symptoms are mainly related to slowly progressing increased intracranial pressure. Computerized tomography and MR imaging are the main diagnostic tests and are capable of leading to the correct preoperative diagnosis in most cases. However, stereotactic biopsy may sometimes be necessary to rule out intraaxial lesions. Surgery should be performed with the aim of obtaining as radical a removal as possible. However, fragments of the capsule should be left in situ if their removal may jeopardize the deep veins to which they adhere. A wise, extensive subtotal removal of tumor may ensure a very long symptom-free interval before an eventual recurrence will become clinically relevant.[1] It is not unusual that reoperation will never be needed to treat epidermoids managed with this treatment philosophy.

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