Radiographic identification and surgical excision of an epidermoid tumor of the pineal gland

Case report

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An epidermoid tumor of the pineal gland for 5 years was incorrectly assumed to be a surgically inaccessible lesion. The patient showed progressive neurological deterioration despite two courses of irradiation therapy and numerous attempts at cerebrospinal fluid shunting. A ventriculogram ultimately demonstrated the epidermoid tumor. Significant improvement resulted from resection of the tumor and the use of a shunting system capable of passing particulatey contaminated ventricular fluid.

KEY WORDS: epidermoid - pineal - ventriculogram - third ventricle - cerebrospinal fluid shunts

Tumors originating in the pineal gland, irrespective of tissue type, are relatively infrequent. Their incidence in American and European brain tumor series is less than 1%; the frequency in Japanese series has been reported as high as 5%. Tumors of the pineal region have been classified by Russell and Rubinstein into four principal categories: teratomas, pinealomas, glial tumors, and epidermoid or pearly tumors. Tumors qualifying for inclusion in the latter category are distinct rarities. We found only seven unequivocal cases of pineal epidermoid tumors in an extensive search of the literature, and each of these cases had a fatal outcome. The case we are reporting appears to be the first published instance of a successful surgical attack on a tumor of this type in the pineal region.

Case Report

A 24-year-old woman was first admitted to the Colorado General Hospital in September, 1964, because of recurrent headache, blurred vision, and formed visual hallucinations. She was lethargic with slowly reactive pupils, severe papilledema, but no gaze palsy. She had been hospitalized for psychiatric reasons 2 years previously. Ventriculography at this time demonstrated only generalized hydrocephalus due to a mass invaginating the third ventricle from the pineal area. Pneumoencephalography was not done. No specific radiographic diagnosis was considered possible (Fig. 1). A ventriculojugular shunt was inserted, and the pineal region irradiated with 5000 R over a 6-week period.
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The patient had been readmitted 2 weeks after discharge because of increased intracranial pressure, relieved by revision of an obstructed shunting device. In April, 1966, she was readmitted because of blurred vision, headache, and paralysis of upward gaze. An obstructed shunt was again revised. She was again admitted to the hospital in July, 1966, because of papilledema and lethargy. A brachial arteriogram demonstrated hydrocephalus, marked upward displacement of the internal cerebral vein, and a mass in the right thalamic area. The ventriculojugular shunt was again revised with subsequent improvement.

The patient was rehospitalized in January, 1968, because of headache, lethargy, and progressive dementia. Cerebral angiography was again performed (Fig. 2). No significant change in ventricular size was noted but increased elevation of the internal cerebral vein indicated continued growth of the lesion. She was given another course of radiation therapy to the pineal region (3000 R over a 6-week period).

Examination. In August, 1968, the patient was rehospitalized because of headache, poor sense of balance, and left-sided weakness. She was lethargic but at times fully oriented. Examination revealed a left hemiparesis, chronic papilledema, complete Parinaud's syndrome with unreactive pupils, and a left sixth nerve paresis. Scalp palpation of the shunting device suggested blockage of the proximal catheter. Right carotid arteriography demonstrated progression of hydrocephalus in addition to further upward bowing of the internal cerebral vein. Despite revision of the obstructed shunt, progressive neurological deterioration ensued. A ventriculogram was performed while she was semicомatose, outlining a large transtentorial mass lesion appearing to communicate with the posterior third ventricle (Fig. 3). This irregularly surfaced tumor mass extended bilaterally but predominantly into the right thalamus, and was diagnosed preopera-

Fig. 1. Ventriculograms, September, 1964, with the head brow down (left) and semi-hanging (right). This unmatched pair of films demonstrates the best identification of the fungating pineal area lesion (arrows) which is invaginating and occluding the posterosuperior third of the lumen of the third ventricle. Supratentorial obstructive hydrocephalus resulted. Direct extension of the lesion is enlarging the right thalamus. Opacification of the fourth ventricle and cisterns was not performed.

Fig. 2. Right common carotid angiogram, lateral projection, venous phase, January, 1968. Diffuse superior bowing deformity of the internal cerebral vein indicates a progressive infravenous mass (open arrows).
Fig. 3. Ventriculogram, August, 1968, hanging head position. Grade 1.5/4 symmetric supratentorial hydrocephalus indicates the probability of shunt malfunction. A large transtentorial mass (arrow) communicating with the dorso-caudal lumen of the third ventricle, and thus containing air, extends in the midline deeply into the rostral vermis. The heterogenous flocculent character of the content of this lesion, with a smooth margin (internal surface of capsule) is very suggestive of epidermoid tumor. Note also the eccentric excursion of the supratentorial component into the right thalamus. Subsequent translumbar pneumography showed no additional connections to the lumen of the lesion, but did demonstrate total obstruction of the caudal aqueduct to retrograde flow of air, and a rather caudal position for the fourth ventricle (see Fig. 4).

tively as an epidermoid or pearly tumor. Shortly after the ventriculogram she became decerebrate and a craniotomy was performed on an emergency basis.

Operation. The pineal region was approached through a right parietooccipital craniotomy with partial excision of the right occipital lobe. The tentorium was incised and retracted, exposing a diaphanous tumor capsule harboring air bubbles and a lobulated tumor mass with a typical mother-of-pearl luster. Resection of the tumor was ac-
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Fig. 4. Epidermoid tumor (non-shafted arrows), illustrated by a midsagittal drawing, reconstructed from pneumographic and surgical observations. Note the superior bowing of the internal cerebral vein over the neoplasm (superior arrow). The lateral ventricles are indicated by dotted outline. Free communication with the lumen of the third ventricle is present (shafted arrow). The aqueduct is totally occluded at both rostral and caudal ends (open arrows).

complished in piece-meal fashion with the exception of small fragments of tenaciously adherent capsule along the distended third ventricular walls. Inspection of the tumor bed after evacuation of the intracapsular contents revealed that the neoplasm had attenuated the midbrain tectum and superior medullary velum, entered the third ventricle to within 1 cm of the foramina of Munro, and indented the right pulvinar (Fig. 4). The fourth ventricle was not entered during the course of the dissection, but the superior vermis was markedly displaced allowing easy visualization of the right superior cerebellar artery.

Microscopic examination of the tumor revealed laminated, cornified tissue with cholesterol crystals, without hair or lining epithelium. The contents were typical of a cholesteatoma or epidermoid tumor. 

Postoperative Course. Convalescence was complicated by poor healing of the previously irradiated scalp, and closure was eventually accomplished by rotation of the full thickness pedicle flap. The patient was maintained on steroids during the surgical procedure and for 6 weeks postoperatively. She became progressively more alert, but 2 months postoperatively suddenly complained of headache and became lethargic. A pneumoencephalogram demonstrated complete occlusion of the aqueduct, and a Denver shunt was implanted as a ventriculoperitoneal diversion. Ventricular fluid protein was 80 mg% at the time of insertion with particulate contamination. Steady neurological improvement has occurred since this procedure, and no further shunt revisions have been required. Sixteen months postoperatively the only significant neurological defi-
cits are a left homonymous hemianopsia and paralysis of upward gaze. She is self-sufficient and cares for her two small children.

**Discussion**

Smaltino and Cuciniello\(^\text{14}\) have recently reviewed the subject of epidermoid tumors situated in the pineal region, and called attention to the feasibility of direct surgical attack. Other neurosurgeons have questioned the conservative measures of initial irradiation and shunting procedures for pineal tumors, and have advocated direct surgical exploration to avoid the tragedy of overlooking a benign lesion.\(^\text{6,8,12,15}\) This attitude must be tempered, however, by consideration of the formidable morbidity and mortality associated with the procedure, as well as the undeniable low frequency of lesions amenable to resection. The epidermoid or pearly tumor is avascular and under optimal conditions can be resected by using current operative techniques. The incidence of pearly tumors in the pineal region is truly “uncommonly rare.”\(^\text{12}\) The more common pineal lesions, such as teratoma or pinealoma, may infiltrate or share critical vascularity with vital mesencephalic structures, thus posing great operative hazard.

This case illustrates that the therapeutic strategy for pineal tumors must be based on comprehensive neuroradiological examination. Pallas, *et al.*,\(^\text{11}\) have emphasized the importance of thorough radiological examination of the posterior third ventricle in planning therapy for pineal tumors. The air-contrast configuration within a pearly tumor, when air fortuitously enters the capsule, is almost pathognomonic for this specific tumor type.\(^\text{1}\) It is noteworthy that air-contrast study was finally definitive in this case, whereas arteriography, although depicting extension into the third ventricle and the progression of hydrocephalus, failed to hint the nature of the lesion.

Retrospective consideration of the dilating, fungating, intraluminal lesion of the posterosuperior third ventricle suggested by studies in September, 1964 is appropriate. At surgical exposure in 1968 this epidermoid tumor had a smooth external capsular surface posterosuperiorly. We conclude that the interface between the neoplasm and the third ventricular lumen, if it ever existed, was also smooth. Therefore, the mottled air surface seen in 1968 must have represented communication through the capsule from the third ventricle into the lumen of the neoplasm. Could this lesion have been diagnosed then as a probable epidermoid tumor? Most pineal area neoplasms present a smooth invagination into the third ventricle, although hypothalamic infiltration might make this somewhat irregular. Occasional neoplasms, however, especially a luminal ependymoma, could have this frond-like configuration. We believe, therefore, that consideration of an epidermoid tumor could only be justified if the ventriculogram showed that air had passed beyond the limits of the lumen of a normal third ventricle. The quality of our emergency ventriculogram in 1964 did not permit this differentiation between “luminal” and extraluminal air passage. Moreover, we now believe that a post-shunting pneumoencephalogram would have been advisable in 1964; it would have been important to know the character of the quadrigeminal cisternal component of the lesion, especially if a smooth surface had been found.

The recurrent failure of shunting devices to control hydrocephalus in this case may have been due to shedding of tumor contents into ventricular fluid. This factor may have been responsible for the acute aqueductal occlusion noted postoperatively. In this regard, the Denver shunt, capable of passing fluid with high protein or particulate contamination, has functioned without difficulty for over 14 months.\(^\text{7}\)

In summary, careful evaluations of the ventricles, aqueduct, cisterns, and lumen of the lesion are essential in the preoperative evaluation of tumors in the pineal region.

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