Epidermoid tumour (tumor perlée, cholesteatoma) of the fourth ventricle: case report and review of literature

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SYNOPSIS The history, clinical course, and the 17 year follow-up after surgery are reported in a patient who had an epidermoid tumour removed from the fourth ventricle. The gross appearance of the contents of the tumour showed multiple tissue aggregates, with the classical pearl-like appearance. Previous cases reported in the literature are collated.

Epidermoid tumours (cholesteatomas) are uncommon intradural intracranial lesions. Cushing (1932) reported 15 cases of epidermoid and dermoid tumours in a series of 2,023 verified brain tumours. One epidermoid involved the fourth ventricle and the upper spinal canal.

Dandy (1966) reported nine intradural epidermoid tumours: four in the cerebellopontine

angle and one each in the tip of the temporal lobe, third ventricle, fourth ventricle, pineal, and sagittal area.

Grant and Austin (1950) reviewed the literature in 1950 and reported 22 epidermoid tumours: 15 were intradural, four in the cerebellopontine angle, four paraspinal, two in the third ventricle, two in the cerebral hemisphere, two in the

TABLE
REPORTED EPIDERMOID TUMOURS OF FOURTH VENTRICLE INCLUDING AUTHOR'S CASE

<i>Author</i>	<i>Total no. cranial and intracranial</i>	<i>Total no. 4th V.</i>	<i>Results after surgery</i>	<i>Duration of follow-up</i>	<i>Comments</i>
Cushing (1932)	15	1	—	—	Tumour extended into upper spinal canal
Mahoney (1936)	112	15	—	—	—
Sheldon <i>et al.</i> (1953)	1	1	Recov.	1 Yr	Pneumoenceph. showed classical filigree lacework pattern
Tytus and Pennybacker (1956)	41*	2†	Recov. (1)	—	* Included also dermoids † Both involv. vermis and 4th V. one vis. by air showed classical filigree lacework pattern
Rosenbluth and Lichtenstein (1956)	2	1	Recov.	19 Mo.	—
MacCarty <i>et al.</i> (1959)	24	1	Died*	—	* Had stupor preop. with bilat. choked disc
David <i>et al.</i> (1965)	1	1	Recov.	15 Da.	Lipoidal ventriculography gave appearance of tumour similar to that with air
Patriquin <i>et al.</i> (1969)	4	4	—	—	One tumour vis. by air showed classical filigree lacework pattern
Dandy (1966)	9	1	—	—	Described these lesions 'The most beautiful tumors in the body'
Pepus <i>et al.</i> (1968)	1	1	Died	—	Tumour mainly cystic
Navarro Ariles <i>et al.</i> (1972)	1	1	Recov.	4 Mo.	—
Fredy <i>et al.</i> (1972)	2	1	Recov.	—	—
Scott (1974)	13	1	Recov.	17 Yr	—

cerebellar hemisphere, and one in the vermis. None were in the fourth ventricle. However, they cited Mahoney (1936) who collected 142 epidermoid tumours from the literature: 112 were intradural and were located as follows—cerebello-pontine angle (53), parasagittal (44), the fourth ventricle (15). Additional cases of epidermoid tumours in the fourth ventricle have been reported by others (Table).

David *et al.* (1965) state that epidermoid tumours of the fourth ventricle comprise approximately 5% of all epidermoids of the cranium and brain.

The classical appearance of this tumour in the lateral ventricles as seen in pneumoencephalography has been reported as diagnostic of an epidermoid tumour by many observers (Dyke and Davidoff, 1937; Hauser and Elkins, 1949; Lindgren, 1952; Scott, 1957). These encephalographic patterns have been described as irregular streaks, filigree lacework of communicating channels of air, reticular masses of air, and

sponge-like irregular collections of air (Dyke and Davidoff, 1937; Lindgren, 1952). This pattern is explained by distribution of the air in the folds of the walls of the tumour and no other brain lesion produces a similar picture (Lindgren, 1952). To the best of my knowledge, no one has reported this pattern in an air study of an epidermoid in the third ventricle. Patriquin *et al.* (1969), Tytus and Pennybacker (1956), and Sheldon *et al.* (1953) have reported that the pneumoencephalographic appearance of the epidermoid tumour in the fourth ventricle is similar to that reported in the lateral ventricles.

Most pathologists agree that the tumour develops from embryonic inclusions of the epidermal portion of the epiblasts. These inclusions may occur anywhere along the base of the brain from the chiasm to the posterior cistern or may involve the cerebellopontine angle. The extradural lesion may appear in the cranial bones, under the temporal lobes, about the bony orbits, or within the middle ear. The interior of the

FIG. 1. Gross appearance of tumour tissue after intracapsular removal. Aggregations of tissue show the classical pearl-like appearance.

FIG. 2. The tumour shows a cyst lining which is composed of flattened squamous epithelium. There are masses of necrotic keratin and a few small aggregates of squamous cells. *H and E*, $\times 178$.

tumour is entirely avascular and resembles 'cottage cheese' in colour and consistency.

Bailey (1948) described the histological structure as follows: 'On the outside is a thin fibrous layer within which are flattened epithelial cells which sometimes contains granules of keratohyaline. The central portion is composed of what one might call the ghosts of epithelial cells; their internal structures have disappeared leaving only the cells. This portion resembles the appearance of a dead plant. Besides the skeletons and debris of dead cells one finds some fat and usually crystals of cholestrin'.

CASE REPORT

G.B., aged 38 years, was admitted to Temple University Hospital on 1 July 1950. She complained of a staggering gait, double vision, vomiting, and suboccipital headaches of six weeks' duration. Her past history was otherwise negative.

The neurological examination showed an alert patient. Her pupils were normal. There was a slow

coarse nystagmus on lateral gaze to either side. Corneal sensation was normal. The visual fields were normal and the eyegrounds showed bilateral choked discs of 3-4 dioptries. Facial power, sensation, and hearing were not impaired. Her tongue was protruded in the midline and the soft palate moved normally.

The deep tendon reflexes were hyperactive bilaterally. There was no Hoffman or Babinski sign. There was no impairment of sensation or strength in the extremities.

The patient was ataxic and veered to the left on walking. There was marked dysmetria, ataxia, rebound phenomenon, and hypotonia in the left upper extremity, with only slight dysmetria and ataxia on finger to nose test on the right. There was marked swaying in the Romberg position.

The provisional diagnosis was a brain tumour in the left cerebellar hemisphere. Radiographs of the skull and chest were normal.

A suboccipital craniectomy revealed an encapsulated, lobulated, plum-sized, glistening, white tumour in a markedly enlarged fourth ventricle. The encapsulated mass was adherent to the floor of the

ventricle. An intracapsular removal was done. The contents had the appearance of a cholesteatomatous tumour. The white tissue appeared flaky, interspersed with areas of 'mother of pearl', and with small round globules of various size, which had the classical appearance of a pearl strikingly demonstrating the appropriateness of the original description 'tumor perlée' by Cruveilhier (1829) (Fig. 1). The capsule was then completely removed from the ventricle, which measured 5 cm in transverse diameter. The dilated rostral portion of the aqueduct of Sylvius was visualized. A silver clip was placed in the roof of the ventricle for future radiological identification. The dura mater was left open and the incision closed.

HISTOPATHOLOGY The tumour tissue showed a cyst lining which was composed of flattened squamous epithelium. There were masses of necrotic keratin and a few small aggregations of squamous cells (Fig. 2). Epidermoid tumour (cholesteatoma) was diagnosed.¹

CLINICAL COURSE The patient made an uneventful recovery. An examination in 1961, 10 years after her operation, revealed no headaches or visual complaints. The decompression area was soft. Her pupils reacted well to light and accommodation. There was no papilloedema and no nystagmus. She was ataxic and tended to veer to the left on walking.

The patient was contacted by letter in May 1967, 17 years after surgery. She had no headaches and no cranial nerve complaints, but she was unable to do her housework because of confinement to a wheelchair for the past six years because of a tremor of the right hand, and a marked ataxia on attempting to walk.

¹ Dandy (1966) believed that the 'cholesteatoma' is a misnomer because cholesterol crystals are not always in these tumours.

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