

A clinicopathological study of intracranial epidermoid cyst.

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Six patients with intracranial epidermoid cysts were reviewed. There were four cerebellopontine angle (CPA) lesions and two pineal epidermoid cysts. There was no sex predilection and the average age was 34.5 years. Headache and cerebellar signs were common among our patients with CPA masses. Sudden onset of hemiparesis was noted in one case due to the associated pontine infarction. Such associated disease is uncommon. One patient with a pineal epidermoid cyst was asymptomatic and appeared unique. CT appearance generally showed either a hypodense or an isodense area. Rim enhancement was noted in only one lesion. There were no fatalities.

Key words: Epidermoid cyst, Pineal epidermoid cyst, Cerebellopontine angle.

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ได้ทำการศึกษาผู้ป่วยด้วยอหิวาต์เฉียบพลันในกะโหลกศีรษะจำนวน 6 รายในช่วง 10 ปี พบว่า 4 รายเกิดที่บริเวณแอ่งระหว่างสมองเล็กและพอนส์ อีก 2 รายที่ต่อมไพเนียล พบในหญิงเท่ากับชาย โดยมีอายุเฉลี่ย 34.5 ปี อาการปวดศีรษะและอาการของสมองเล็กมักพบในผู้ป่วยกลุ่มแรก ผู้ป่วยรายหนึ่งมีหอย่อมตายของสมองบริเวณพอนส์ร่วมไปด้วย ผู้ป่วยอีกรายตรวจพบก้อนที่ต่อมไพเนียลโดยบังเอิญด้วยเครื่องเอกซเรย์คอมพิวเตอร์ ลักษณะที่ตรวจพบโดยรังสีวิทยาด้วยเครื่องเอกซเรย์คอมพิวเตอร์มักเป็นก้อนที่มีความหนาแน่นต่ำหรือปกติ ไม่พบผู้ป่วยที่ถึงแก่กรรมภายหลังผ่าตัด

Generally intracranial epidermoid cysts are slow growing benign tumors, although malignant transformation has been occasionally recorded.⁽¹⁻³⁾ They comprise about 0.2-1 per cent of all intracranial tumors.⁽⁴⁾ The common sites affected by this tumor are the cerebellopontine angle (CPA), parapituitary, diploe, rhomboid fossa, and the intraspinal regions. It has occasionally been found in the brain parenchyma and the pineal.^(4,5) Six cases of intracranial epidermoid cysts (four CPA lesions and 2 pineal masses) are described here. One example is an asymptomatic pineal epidermoid cyst which is unusual in our experience.

Materials and Methods

Six cases of intracranial epidermoid cysts were obtained from the Department of Pathology, Chulalongkorn Hospital over a 10-year period (1984-1994). The tissues were fixed in 10 per cent formalin and embedded in parafin. Sections were stained with hematoxylin and eosin (H&E). All clinical records of these subjects were studied after review of the microscopic materials.

Results

The clinical features are given in Table 1. There were 3 men and 3 women. The youngest patient was a 27-year-old woman and the oldest patient was a 45-year-old man. The average age was 34.5 years. The symptomatic onset ranged from 1 month to 2 years. The majority of patients had a short duration of illness, less than 1 year. There were four cases located in the CPA and two in the pineal. Among the patients with CPA lesions, three had progressive headache and cerebellar signs. Papilledema, hearing impairment and cranial nerve palsies were each noted in only one case. Only one case was presented with headache and left hemiparesis, and this was due to the associated right pontine infarction. Regarding patients with pineal masses, one patient had only progressive headaches and bilateral papilledema, the other was asymptomatic. The latter patient was a 45-year-old man who had been invited to a computer tomography (CT) scan demonstration. A demonstration scan on him incidentally revealed a pineal cyst. CT scans 2 and 4 months later revealed a slow progressive enlargement of the lesion. At craniotomy a 2×1 cm well circumscribed cyst was totally removed. The patient was neurologically normal when he was examined 1 year after surgery.

Table 1. Clinical and pathological features in 6 cases of intracranial epidermoid cysts.

Case No.	Age	Sex	Location	Duration	Size (cm)	Clinical features	CT appearances	Pathological findings
1	27	F	CPA	2 yr.	5×5	Cerebellar ataxia, dysphagia, dysarthria, deviation of tongue to the right	Hypodense with calcification, and hydrocephalus	Foci of calcification
2	28	F	CPA	7 mo.	7.5×5	Headache, papilledema, cerebellar ataxia	Hypodense with rim enhancement, hydrocephalus	No calcification
3	31	M	CPA	1 mo.	1.5×1	Headache with L. hemiparesis (1 wk)	Hypodense with hydrocephalus, R. pontine infarction	No calcification
4	38	M	CPA	1 mo.	Large	Headache, L. hearing loss cerebellar ataxia	Isodense with hydrocephalus	No calcification, mild gliosis
5	38	F	Pineal	1 yr.	2.5×2	Headache, papilledema	Hypodense with hydrocephalus	Focal calcification
6	45	M	Pineal	-	2×1	Incidental finding	Hypodense with hydrocephalus	No calcification

M= male; F = female; yr = year; mo = month; L = left; R = right.

Concerning the radiographic findings, CT scans were performed in all cases. A hypodensity mass was noted in five cases and an isodensity mass in one example. Rim enhancement after contrast injection was demonstrated in one instance (case 2). Calcification was noted in case 1. CT also showed hydrocephalus in all patients.

Pathologically, the cystic lesions ranged from 1.5 cm to 7.5 cm and contained soft white and waxy material. Calcification was grossly visible in

case 1. Microscopically, the lining of the cysts was stratified squamous epithelium resembling skin but no other skin appendages were seen (Figures 1A, 1B). The content was a layer of keratinous material (Figure 1B). Foci of calcification were found in two lesions (cases 1, 5). Additionally, vascular proliferation in the adjacent tissue around the cyst was noted in case 2 (Figure 2). No evidence of malignancy or granulomatous inflammation was observed.

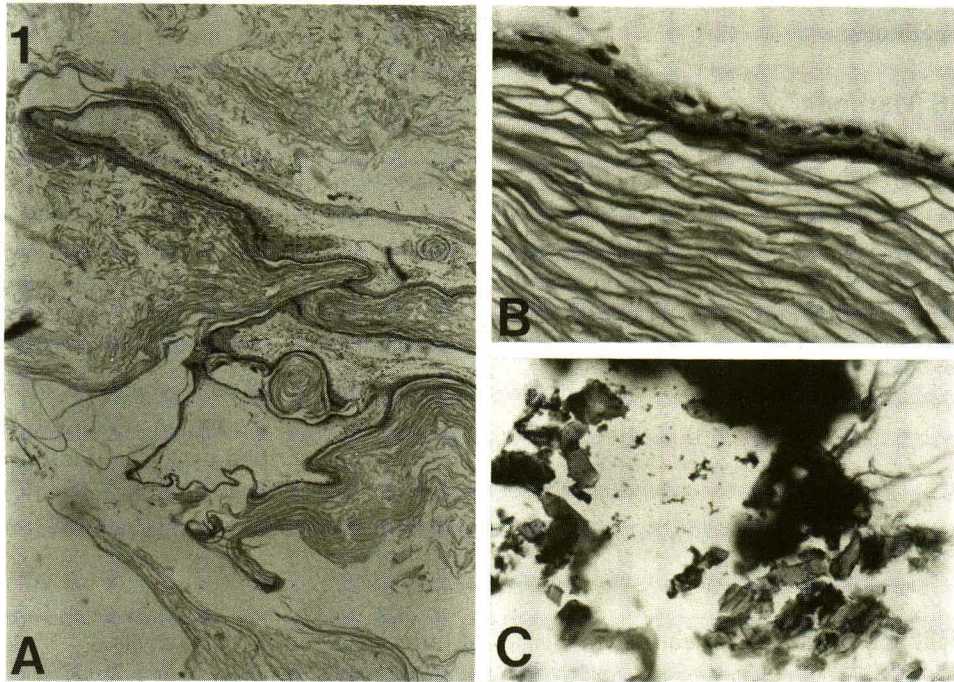


Figure 1. Photomicrographs of intracranial epidermoid tumor

A. Thin-walled cyst with desquamative lamellae. (H&E \times 100)

B. Higher-power view showing mature stratified squamous epithelium without skin appendages. (H&E \times 250)

C. Calcification within the wall. (H&E \times 250)

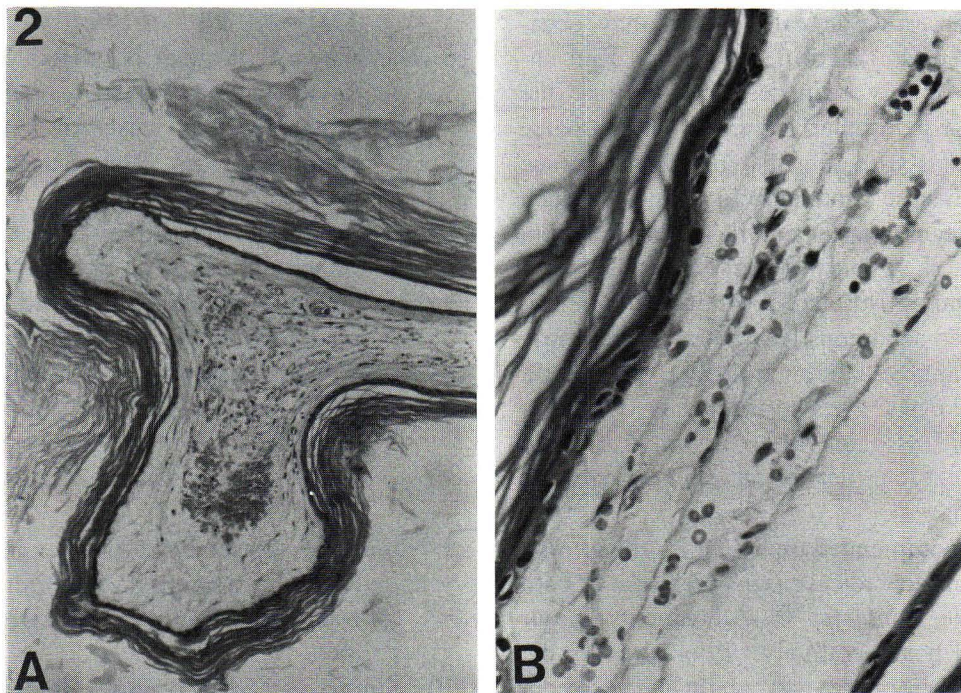


Figure 2. Microscopic features of the lesion in case 2.
A. Cystic wall with vascular spaces. (H&E x 100)
B. Numerous thin-walled vessels. (H&E x 250)

All patients underwent surgical excision. Only one had facial palsy postoperatively which required facial-hypoglossal anastomosis (case 1). Another patient developed epidural hematoma and persistent hydrocephalus which required a second craniotomy and ventriculoperitoneal shunt (case 5). There were no fatalities.

Discussion

The age distribution of intracranial epidermoid cysts is wide ranging from birth to 80 years, but the peak onset is in the fifth decade.^(2,4) Our study revealed that the tumors tend to become symptomatic between the age of 20 and 40 years. According to Baxter and Netsky,⁽⁶⁾ the lesions occur more often in men while in some series women are predominant.⁽²⁾ Our data showed no sex predilection but our sample was small. The correlation between the size of the tumors and the duration of illness was not clearly demonstrated in this study due to the limited number of patients. However, it is still reasonable to suggest that tumors larger than 2.5 cm were found mostly in patients with less than 1 year of symptoms (Table). The tumor may remain quiescent, as noted in one of our cases (case 6).

Headache, cerebellar signs, and increased intracranial pressure were frequent symptoms in our patients with CPA lesions, similar to most other reported cases.^(6,7) Most patients in our study had at least two symptoms. Such clinical manifestations are, of course, attributed to the mass effects caused by a space-occupying lesion. The preoperative evidence of right pontine infarction due to atherosclerosis in one of our patients is uncommon and we are not aware of its previous description.

Epidermoid cysts in the pineal region are even rarer than for the CPA lesions.⁽⁵⁾ Kirsch and Stears in 1970 collected only 7 cases of such tumor descriptions in the literature, all with fatal outcomes. The authors also added an additional case which was successfully treated surgically.⁽⁸⁾ As far as can be ascertained, only 18 cases of pineal epidermoid cysts including the two current cases have been recorded.^(5,8-14) The patients often had headaches, intracranial hypertension, hydrocephalus, visual impairment, altered consciousness, and seizures. The incidental finding of such a pineal lesion in the absence of clinical manifestations in one of our patients (case 6) appears unique.

CT scan is a valuable tool to identify the lesions. Characteristically, the tumors appear as

hypodense masses while calcification may occasionally be seen, as noted in our series. Rim enhancement has rarely been described because the tumors are poorly vascularized.⁽¹⁵⁻¹⁸⁾ The presence of many vascular spaces in the surrounding tissue as noted in our case 2 probably accounts for such unusual enhancement after contrast administration. Pathologically, the typical solitary mass is pearly white with a smooth surface. The microscopic findings of stratified squamous lining with a concentric layer of desquamated keratin are diagnostic criteria for epidermoid cysts.^(2,4) The absence of other skin appendages are also useful to distinguish these tumors from other similar lesions such as dermoid cyst or teratoma.^(2,4)

The recommended treatment for this benign intracranial tumor is radical surgical removal of both the cyst and its contents.⁽¹⁹⁾ All patients in our series did well postoperatively. Prior to 1936, the operative mortality rate was about 70 per cent but this dropped to less than 10 per cent in the 1970's.⁽¹⁹⁾ In a recent report, Yasargil et al, encountered no fatalities in 35 cases of intracranial epidermoid cyst removal by utilizing microsurgical techniques.⁽⁷⁾ The greatly improved mortality rate is undoubtedly the result of improved surgical techniques, neuroradiology, and neuroanesthesiology as was noted in our study and in others.^(6,7,19)

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