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Benign Symptomatic Lesions of the Pineal Gland. Report of Seven Cases Treated Surgically

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Summary

Seven patients with non-neoplastic symptomatic lesions of the pineal gland have been operated on in our department since 1988. Clinical symptoms were slight and there were intermittent signs of a space occupying lesion in the quadrigeminal area, i.e. visual and gait disturbances. In three patients, obstructive hydrocephalus with related symptoms was found. Diagnosis was confirmed by use of MRI in sagittal planes, revealing a cystic lesion in five and a solid tumour in two patients. All patients were operated on without complications using an infratentorial supracerebellar approach. Histological examination showed glial cysts of the pineal gland in five patients. In the two solid specimens, normal pineal tissue was found. These lesions seem to be of special interest, as the only pathological property are their size: Both lesions – "too large pineal glands" – caused obstruction of the outlet of the third ventricle with subsequent hydrocephalus. Surgical treatment was curative in all cases with prompt relief of the symptoms. Clinical symptoms and signs, diagnostic and pathological findings, as well as the surgical results of these cases will be reported.

Keywords: Pineal cyst; enlarged pineal gland; infratentorial supracerebellar approach; MRI; occlusive hydrocephalus.

Introduction

Symptomatic non-neoplastic lesions of the pineal gland are rare. The first descriptions of two cases with cystic enlargement were given by Campbell in 1899[2]. In 1914, Pussep published the case of a 10 years old boy suffering from progressive oculomotor dysfunction and ataxia, followed by symptoms of hydrocephalus with headache, drowsiness and optic atrophy[17]. Clinically, he suspected a tumour of the quadrigeminal region and performed surgery by use of a supracerebellar approach. The operative finding was a cystic lesion of the pineal gland, which could be confirmed by autopsy following death of the patient three days after surgery. Following this case, only occasional reports concerning cystic lesions of the pineal gland were published by different authors, but diagnosis of this entity remained very rare[1, 3, 4, 9, 18]. Evidently, this changed after the introduction of MRI. Especially the direct high resolution imaging in sagittal planes enables a clear view of the pineal region. In this context, Lee and co-authors (1987) found 15 cases of pineal cysts among a total number of 1000 MR examinations, of whom two patients underwent surgery[13]. Recently, Klein and Rubinstein (1989) reported seven cases of symptomatic glial cysts of the pineal gland referred to them for consultation on the histological diagnosis[10]. Since 1988, we have observed seven patients with non-proliferative symptomatic lesions of the pineal gland, who have been operated on.

Patient Material, Diagnostic and Surgical Methods

Clinical data, MR-findings and histology of the lesions are summarized in Table 1.

Surgical Treatment

All patients were operated on by the infratentorial supracerebellar approach (Fig. 1), first performed and advocated by Krause in 1913[14] and adapted for microsurgical purposes by Stein in 1971[19] and others[11, 13, 14, 15, 16]. We performed an osteoplastic occipital trepanation, using four burr-holes. The upper two were placed exactly above the transverse sinuses. Neither intraoperative, nor postoperative ventricular drainage nor opening of the foramen magnum or resection of the C_1-lamina were performed. The dura was opened in a tongue-like fashion with the base to the sinuses and was closed skillfully following removal of the lesion. Only the draining veins of the surface of the cerebellum to the tentorium were sacrificed, as well as the vein draining from the upper part of the vermis to the vein of Galen. All other vessels were strictly preserved. In all patients, the lesion was easily accessible and could be clearly separated from the surrounding vessels, as well as from the upper aspect of the

Table 1. *Clinical Symptoms, MR-Findings and Histological Examination Results*

Case °	Age/sex	Symptoms and signs	MR-findings	Histology
1	41/m	headache, drowsiness (acute)	cystic/solid enlarged pineal mass, mild HC	glial cyst
2	30/f	headache, vertigo, gait and visual disturbances (intermittent)	cystic pineal lesion	glial cyst
3	10/m	seizures	cystic pineal lesion	glial cyst
4	19/f	headache, intermittent Parinaud syndrome	cystic pineal lesion + tumour IV. ventricle	glial cyst
5	29/f	vertigo, intermittent visual disturbances	cystic pineal lesion	glial cyst
6	29/m	hydrocephalus	solid pineal mass	no cystic lesion, abundant pineal tissue with calciferous bodies
7	33/m	vertigo, gait and visual disturbances	solid pineal mass + marked HC	

quadrigeminal plate and thalamic walls. Following lesion removal, a wide opening of the third ventricle could be seen.

Case Reports

Two cases will be described in further details:

Case 2

This 30 year-old female, suffering from congenital deafness, had a long history of headache, visual and gait disturbances and vertigo. Encephalitis disseminata or vegetative lability were diagnosed pre-

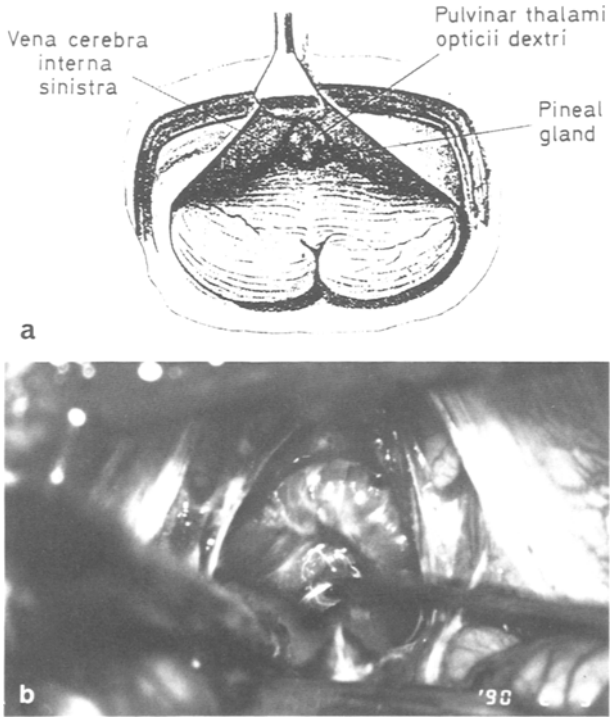

Fig. 1. Surgical approach to cystic lesions of the pineal gland. Above: Schematic drawing of the infratentorial supracerebellar approach (from Pussep's original publication in 1914). Below: Microsurgical operative view of a large pineal cyst in a 30 year-old female (case 2)

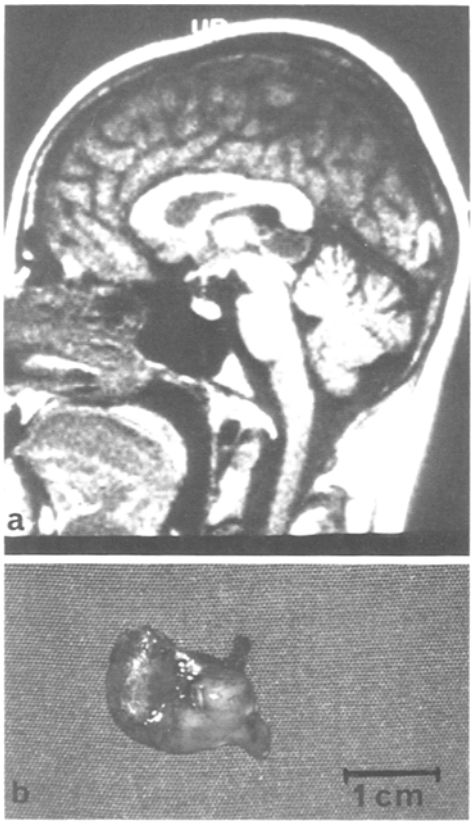

Fig. 2. MRI (above) and surgical specimen (below) in a patient with a large pineal cyst (2,0 cm in diameter); case 1

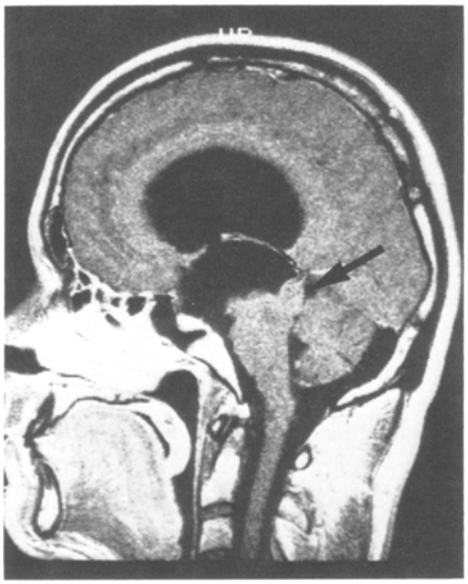

Fig. 3. MRI (midsagittal plane) in a 29 year-old male (case 6), showing marked obstructive hydrocephalus of aqueductal stenosis type, caused by a solid lesion of the pineal gland (arrow)

viously. MRI revealed a cystic, pear-shaped lesion in the pineal region (Fig. 2).

Using the infratentorial/supracerebellar approach (Fig. 1), the cyst was removed completely (Fig. 2). The postoperative course was uneventful. Histologically, a glial cyst was found (Fig. 4 a).

Case 6

This 29 year-old man presented with a one-year history of mild intermittent symptoms with vertigo, ataxia and visual disturbances. Following rapid development of headache and nausea, he was admitted to our department with marked obstructive hydrocephalus. CT scanning suggested an aqueductal stenosis. MRI, however, clearly demonstrated the cause of the obstruction by a solid space-occupying lesion of the pineal gland (Fig. 3). Following a primary shunting procedure and after recovery of the patient, the lesions was removed without complications. The histopathological examination of the surgical specimen revealed pineal tissue with calciferous bodies without any cystic components (Fig. 4 b).

Results

1. Histological Examination

The results of histologic examinations of the pineal lesions are summarized in Table 1. In 5 cases, the di-

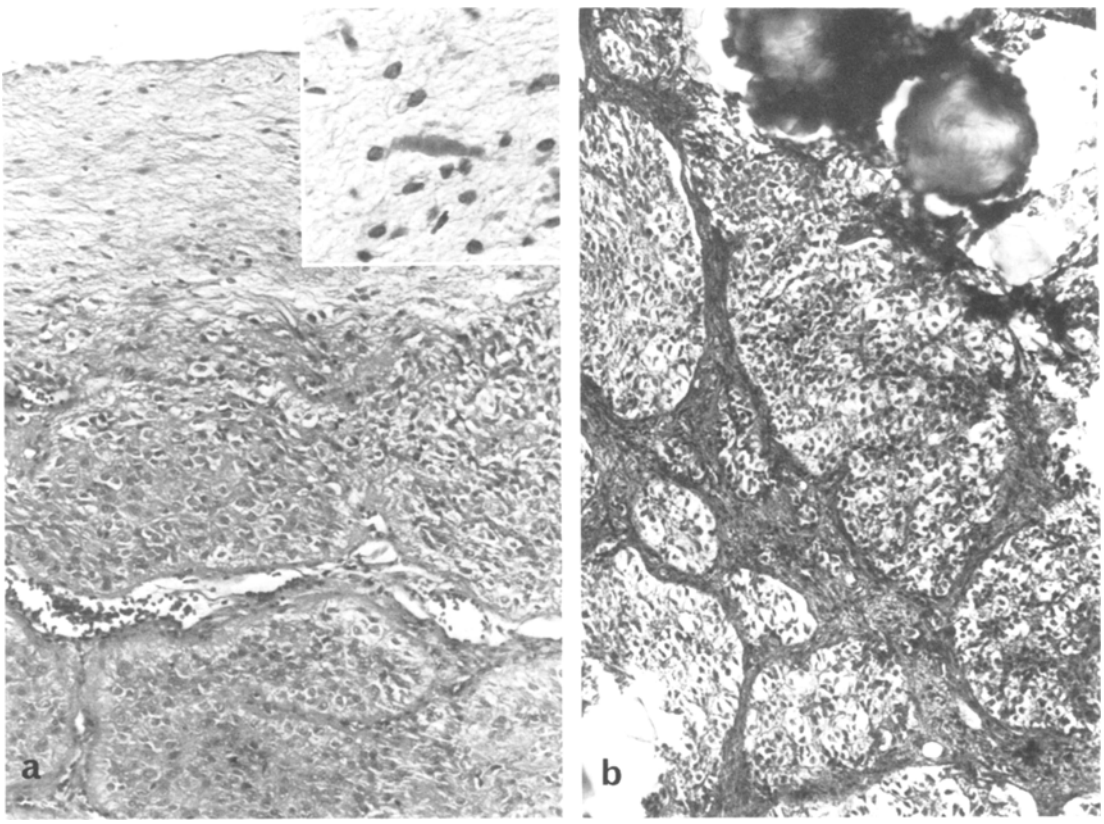

Fig. 4. Histological findings in benign space-occupying pineal lesions. a) Wall of a glial cyst of the pineal body (case 2): Periluminal layer composed of gliotic tissue with Rosenthal fibres (shown in a higher magnification in the inlet); compressed pineal parenchyma in the upper third of the cutout. HE staining, × 140, inlet × 340. b) Pineal tissue with typically lobulated pattern and strains of connective tissue (case 6). Calciferous bodies on top. EvG staining, × 140

agnosis of a glial cyst of the pineal gland was made. The inner layer of the cystic walls was found to be composed of a meshwork of glial fibrils with small nuclei of glial cells. Rosenthal fibres were present in varying numbers. Peripheral to this inner layer, compressed pineal parenchyma with corpora arenacea, small isolated nests of pineal cells, and, not in every case, remnants of a fibrous capsule were found (Fig. 4 a). In two cases typically lobulated pineal tissue with abundant calciferous bodies but no cystic lesions were observed (Fig. 4 b).

2. Results of Treatment

The surgical procedure was tolerated very well in all patients. Remittence to the normal care unit was possible one day following surgery and discharge from the hospital seven days later. Mild visual disturbances occurred in three patients, but discontinued completely during the postoperative course. There were no postoperative complications in this series.

In the follow-up, a complete remission of the clinical symptoms and signs was observed. In case 6, treated first by a shunting procedure, the shunt system could be removed in the meantime.

Discussion

Although symptomatic pineal cysts are known since Campbells publication in 1899, such lesions remained a very rare entity until recently. Whereas small cystic components are very frequently found in autopsy specimens, large cysts – greater than 0,5 cm in diameter – occur only in a small number of patients. Various theories regarding the origin of such large cysts, embryogenetic development failures, ischaemic glial degeneration, invagination effects of the pineal gland or haemorrhagic events have been proposed[10]. Klein and Rubinstein recently discussed hormonal influences related to pregnancy or the menstrual cycle which cause enlargement of pre-existing cystic components and the typical symptoms of a space-occupying lesion in the region of the quadrigeminal plate, as they are described in the classical publications of Pussep[17], Krause[12], Förster[5, 6] and Oppenheim[14]. However, there are some discrepancies in our findings: Following the "ischaemic degeneration theory"[4] marked affliction of elderly patients would be expected. However, mean age in our series was 28 years, similar to the report of Klein and Rubinstein. The same argument is valid against the "invagination theory"[7]. In none of our cases, were any signs of haemorrhage present, thus not supporting the "haemorrhage theory"[8].

Finally, our observations give lend no support to the "hormonal related theory", as four out of the seven patients were male and in none of the female cases, were clinical symptoms correlated in time with the menstrual cycle or a pregnancy.

Summarizing, our clinical, imaging and histological findings none support any of the previously given explanations, nor do any cases show consistent features of a clearly definable origin.

In two cases, no cystic lesions where found, but the surgically removed specimen exhibited a histologically normal pineal gland. The only "pathological" feature causing the symptoms of a space occupying lesion was the size of the gland, which was evidently too large for the individual topographical relations between the splenium of the callosal body, the quadrigeminal plate and the outlet of the third ventricle. Thus both of these patients developed marked hydrocephalus. These pathological relationships could be clearly detected by MR-imaging as shown in the case mentioned above. This entity of the "too large pineal gland" observed in two patients of our series has not been described before. Therefore, further data are necessary to prove a predictive value of this syndrome.

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