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Cystic lesions of the pineal region – MRI and pathology

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Abstract Pineal lesions are rare. Tumours in this location comprise 0.4–1 % of intracranial tumours. They grow mainly as solid-mass lesions, and cystic tumours are not common. On MRI, a cystic configuration is associated usually with non-neoplastic pineal lesions rather than with a tumour, but analysis does not allow cystic pineal tumours to be distinguished from glial cysts with certainty. We compared neuroradiological and pathological data from 13 cystic pineal lesions, analysing preoperative MRI. Formalin-fixed, paraffin-embedded surgical specimens were stained routinely and immunocytochemically, using the streptavidin-biotin-complex method. Histology revealed six pineocytomas, four glial cysts, an arachnoid cyst, a low-grade astrocytoma and a teratoma. Signal characteristics of pineocytomas were similar in many respects to those of glial pineal cysts. Histomorphological analysis allowed unambiguous discrimination between pineocytomas and glial pineal cysts.

Key words Pineal region · Cysts pineal · Pineocytomas · Magnetic resonance imaging

Introduction

Primary intracranial tumours of the pineal region represent 0.5 % of intracranial neoplasms. The majority are germ-cell tumours, gliomas and pineal parenchymal tumours; less than 15 % are rare tumours of various origins. The vast majority of these tumours grow as solid, often infiltrative mass lesions; a cystic appearance is unusual [1]. A cystic appearance is often found with glial pineal lesions such as arachnoid and glial cysts.

Since the introduction of MRI, pineal cysts have been found in 1.4–4.3 % of healthy subjects [2]. They are often asymptomatic. Several groups have regarded them as benign, but the majority of reports do not include histology [2, 3]. We sought to establish a correlation between MRI and histological features by reviewing 13 pineal region cysts biopsied over a 6-year period.

Material and methods

We removed 13 small cystic pineal lesions from young patients, seven male and six female (mean age 10 years) between 1989 and 1995. There were 10 patients who presented with distinct headaches and five who had complex focal seizures. Neurological examination revealed disordered eye movements in three and two

Table 1 Clinical and imaging data. Age, sex, signs and symptoms, hydrocephalus, shape and size (craniocaudal "transversal" anterior-posterior diameter in mm) on MRI and characteristics of gadolineum enhancement

Case	Age (years)	Sex	Symptoms and signs	Hydrocephalus	MRI			Diagnosis
					Shape	Size (mm)	Contrast enhancement	
1	8	M	Tremor, complex partial seizures	No	Ovoid cyst with thick wall	8 × 9 × 7	Cyst wall	Pineocytoma
2	7	F	Headache	No	Ovoid cyst	10 × 8 × 10	Cyst wall	Pineocytoma
3	9	F	Headache, complex partial seizures	No	Septate, eccentric cyst with thick wall	18 × 20 × 15	Cyst all	Pineocytoma
4	7	M	Circadian rhythm disturbance	No	Ovoid cyst with thick wall	10 × 9 × 9	Cyst wall	Pineocytoma
5	10	M	Headache impaired concentration	Yes	Ovoid cyst with dorsal nodule	32 × 28 × 35	Cyst wall and tumour nodule	Pineocytoma
6	7	F	Headache, complex partial seizures	No	Ovoid cyst with thick wall	10 × 8 × 8	Cyst wall	Pineocytoma
7	25	F	Headache, abducens palsy	No	Mixed solid and cystic mass	10 × 15 ×10	Cyst wall, without tumour nodule	Astrocytoma
8	15	M	Headache, parinaud's syndrome	Yes	Lobulated cystic-nodular mass	40 × 30 × 45	Tumour nodules and cyst wall	Mature teratoma
9	24	F	Headache, nausea	No	Ovoid cyst	15 × 12 ×20	Cyst wall	Glial cyst
10	5	M	Headache, complex partial seizures	No	Septate, eccentric localized cyst	15 × 9 × 12	Cyst wall	Glial cyst
11	4	F	Ptosis (suspected myasthenia gravis)	No	Septate cyst	10 × 13 ×10	Cyst wall	Glial cyst
12	4	M	Headache, complex partial seizures	No	Ovoid cyst	5 × 6 × 6	Cyst wall and cyst fluid	Glial cyst
13	7	M	Absences, epilepsy	No	Eccentric localised cyst	10 × 12 ×10	No enhancement	Arachnoid cyst

patients developed signs of hydrocephalus. No patient exhibited postoperative complications.

Preoperative MRI was available for all patients. Axial T2- and proton density-weighted images 5–7 mm thick and T1-weighted images 3–7 mm thick had been obtained with spin-echo sequences at 1.5 T. All patients received gadolinium contrast medium. The surgical specimens were formalin-fixed, paraffin-embedded and stained by haematoxylin and eosin (H & E), Goldner and Gomori silver stains. Immunohistochemistry, using the streptavidinbiotin-peroxidase complex method, was employed with the following antibodies: glial fibrillary acid protein (GFAP), synaptophysin, S-100 protein, neurone-specific enolase (NSE), neurofilament protein (NFP) and chromogranin. MIB-1 was used to determine proliferative activity.

For histological and immunocytochemical controls of pineal morphology we studied 16 pineal glands of all age groups from autopsy cases; none contained any form of cyst or tumour.

Results

We diagnosed six pineocytomas, four glial pineal cysts, one arachnoid cyst, one astrocytoma and one teratoma using the WHO classification (1993). Clinical, radiological and diagnostic data are summarised in Table 1.

Signal characteristics were analysed according to the histological diagnosis as follows.

Pineocytomas

Pineocytomas were diagnosed in patients aged 7–10 years. Five appeared on MRI as small, concentric, cystic lesions 8–20 mm in diameter. One much larger tumour contained a dorsal solid portion with strong contrast enhancement (Fig. 1 a).

The signal characteristics were similar in many respects to those of glial pineal cysts. Irregularities of the cyst wall were observed in five pineocytomas, which gave a nodular contrast enhancement. Histology (Fig. 1 b) and the morphological criteria employed will be discussed below.

Glial pineal cysts

Cysts from four patients (aged 4–25 years) appeared on MRI as rounded, partly separated midline cysts,

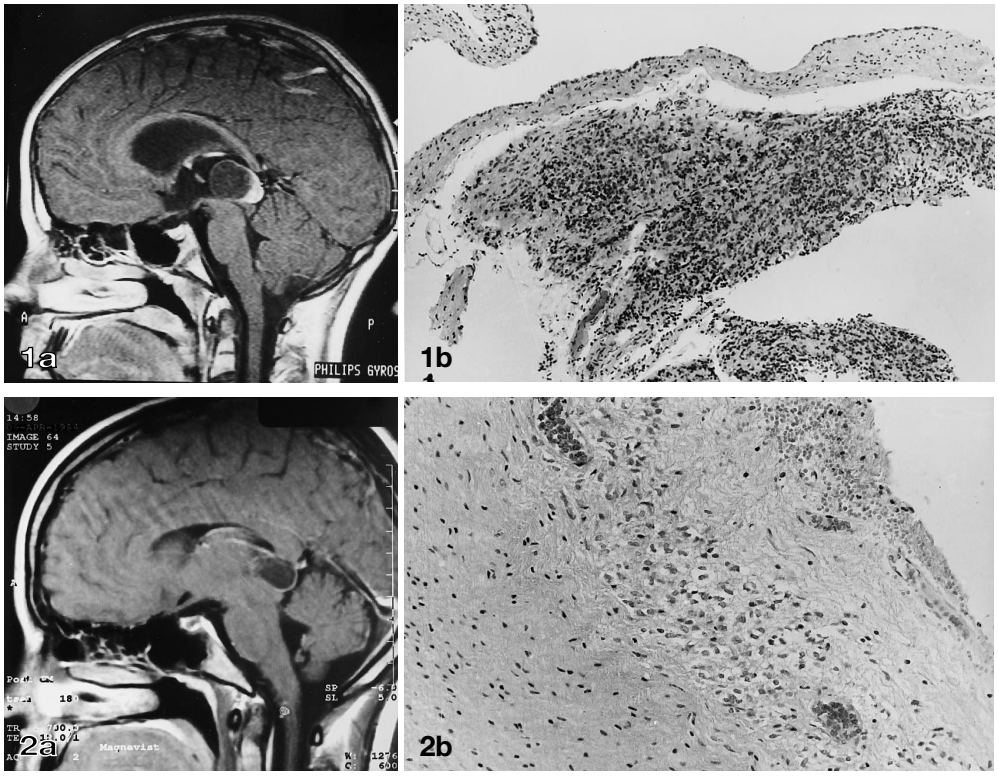

Fig. 1 **a** Cystic tumour with a strongly enhancing solid dorsal component on a sagittal T1-weighted image; there is hydrocephalus. **b** Biopsy shows a well-defined focus of regular small cells with hyperchromatic nuclei, indicating a pineocytoma. Haematoxylin and eosin, original magnification, × 50

Fig. 2 **a** Glial pineal cyst on a contrast-enhanced sagittal T1-weighted image: slight compression of the colliculi and the rostral aqueduct, without hydrocephalus. **b** Typical cyst wall, consisting of an inner gliotic layer with copious Rosenthal fibres *(left),* columns of pineal parenchyma *(middle)* and an outer fibrous capsule *(right).* Goldner, original magnification, × 50

6–20 mm in diameter (Fig. 2 a). In all four patients the well-defined, smooth cyst margin maximum thickness of 2 mm showed moderate, homogeneous contrast enhancement.

Histologically, those cysts showed cyst-wall fragments and residual pineal parenchyma. The wall was composed of three layers – an inner gliotic layer with a high number of Rosenthal fibres, a middle layer with columns of pineal parenchyma and thin fibrous external layer (Fig. 2 b).

Arachnoid cyst

The biopsy material contained small fragments of arachnoid and choroid plexus. An eccentric cystic lesion, 12 mm in diameter, was seen with the MRI signal characteristics of water and no contrast enhancement.

Astrocytoma

Different signals were seen in a 25-year-old woman with a concentric midline tumour, mimicking a pineal cyst on T1-weighted images, but with the T2- and proton density-weighted images showing a solid, low-signal tumour. Histological examination confirmed a cystic low-grade astrocytic tumour with compression of pineal tissue. The majority of the tumour cells expressed GFAP.

Teratoma

A 15-year-old boy showed a large, lobular cystic tumour 45 mm in diameter on MRI. Histology revealed typical features of a teratoma with portions of several germinal layers and expression of mesenchymal, glial and epithelial markers on immunocytochemistry. No adipose tissue was present.

Discussion

We focused on diagnostic criteria for differentiating pineocytomas from glial cysts. In eight cases the diagnosis of a pineal tumour was made; six were pineocytomas. The radiological characteristics of pineocytomas and pineal cysts were usually very similar and characterised by high signal on T2- and only slightly increased signal on T1-weighted images. Glial pineal cysts had no specific features on imaging, but did have thin, smooth walls no more than 2 mm thick, with moderate contrast enhancement. A cyst wall with a maximum thickness of 2 mm has been reported [4–6], and this may be a reliable criterion for the differential diagnosis of

glial pineal cysts from pineocytomas. Irregularities of the wall, with nodular contrast enhancement, suggest a pineocytoma, but this appearance has also been reported in glial cysts [4, 5].

The great diversity of morphological appearances in the tissues of the pineal region may cause serious problems for the histological diagnosis, especially when biopsies are small. Analysing the studies of Fain et al. [4], Borit et al. [7] and Vaquero et al. [8], it appears that the diagnosis of pineocytoma was made when the following criteria were met (Fig. 1 b): a pseudolobular arrangement of small, round cells with hyperchromatic, moderately pleomorphic nuclei and irregular large "pineocytic" rosettes [7]. In addition, neuronal differentiation, with NSE- and synaptophysin-positive cells and variable NFP expression, was seen but there was no proliferative activity. Residual pineal parenchyma showed GFAP-positive cells. In contrast, the histology of pineal glial cysts is well defined (Fig. 2 b) [4–6]. In the present study, pineocytomas could be distinguished from glial cysts with confidence on histological criteria.

The MRI and histological characteristics of the 13 pineal lesions in the present study differ in many respects from previously reported series; they differed in size, neuroimaging appearances and histological pattern, as well as in age, response to therapy and clinical outcome. Few radiological studies contain large numbers of tumours derived from pineal cells [1, 7]; pineocytomas are described as large, solid masses and exclusively cystic tumours are rare. The results of our MRI and histopathological study show contrasting results. In only one case was a large, cystic tumour, with a dorsal solid nodule detected on MRI. In the other five cases, pineocytomas were well defined, cystic lesions 8–20 mm in diameter indistinguishable from glial cysts on MRI, even with contrast medium.

The youth of the patients (7–10 years) with small cystic pineocytomas was also at variance with other studies in which pineocytomas presented as large, solid, often infiltrative tumours, in adult patients. There are few reports on pineocytomas in the first two decades of life [1, 8].

There was a good clinical outcome following operation in all six of our patients with pineocytoma. In other studies, radiotherapy has often been recommended for such patients, even prior to histological confirmation [8, 9]. It has been stressed that in young patients with pineal lesions, one can set aside current therapeutic concepts whereby treatment should depend upon the histological diagnosis. As emphasised in this report, a firm histopathological diagnosis is of primary importance.

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