

# Multiple Epidermoid Cysts Located in the Pineal and Extracranial Regions Treated by Neuroendoscopy

## —Case Report—

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### Abstract

A 22-year-old woman presented with a rare case of multiple epidermoid cysts located in the pineal and extracranial regions. Magnetic resonance (MR) imaging showed a lesion in the pineal region as hypointense on the T<sub>1</sub>-weighted image and hyperintense on the T<sub>2</sub>-weighted image, without enhancement. Neuroendoscopic treatment was performed under a diagnosis of pineal cyst. However, the cyst wall was too thick to perforate, although third ventriculostomy was performed. Diffusion-weighted MR imaging demonstrated the lesions in the pineal and extracranial regions as marked hyperintensity. The diagnosis was epidermoid cyst. Subsequently, neuroendoscopic treatment of the pineal epidermoid cyst was performed. Careful preoperative diagnosis of epidermoid cysts based on diffusion-weighted MR imaging is required.

Key words: pineal region tumor, neuroendoscope, epidermoid cyst

### Introduction

Epidermoid cysts are relatively rare, and account for only 0.2–1% of all intracranial tumors.<sup>11,16)</sup> They are generally found at the base of the skull, in the sellar or parasellar region, the cerebellopontine angle and certain other locations.<sup>2,6,11,15,16)</sup> Epidermoid cysts in the pineal region are very rare,<sup>2,7,8,11,12,16)</sup> and cases with multiple epidermoid cysts located in both the intracranial and extracranial regions are hardly ever reported. We present a rare case of multiple epidermoid cysts in the pineal and extracranial regions.

### Case Report

A 22-year-old woman complained of headache, and was admitted to a local hospital in August 1996. Computed tomography (CT) scan revealed a cystic lesion in the pineal region and associated obstructive hydrocephalus (Fig. 1). CT also showed a low-density area in the superior lateral orbital region. Ventriculoperitoneal shunt placement was performed, following which she was free of headache.

She presented with somnolence and upward gaze

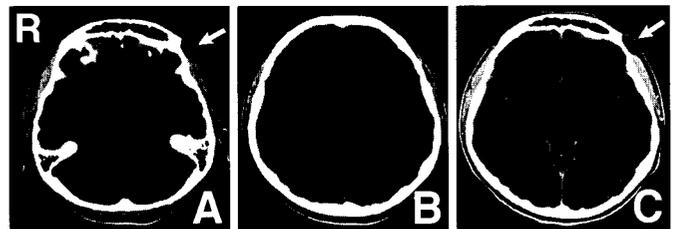
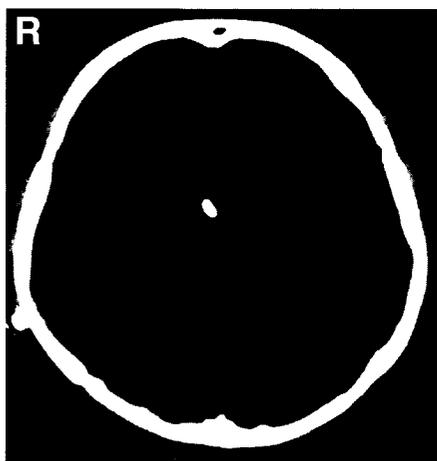


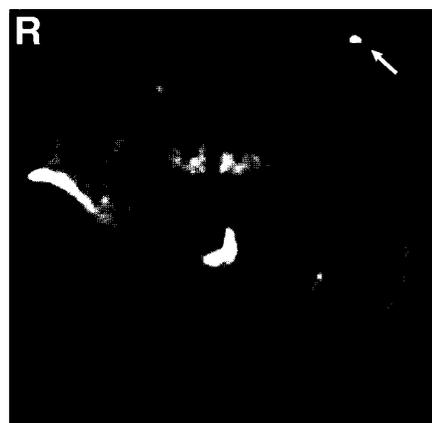
Fig. 1 A, B: Initial computed tomography (CT) scans showing obstructive hydrocephalus due to a cystic mass in the pineal region, and a low-density mass in the superior lateral orbital region (arrow). C: CT scan with contrast medium showing no enhancement of the cystic mass.

palsy with headache in March 1999. CT performed at a local hospital revealed enlargement of the cystic lesion in the pineal region (Fig. 2), as the diameter of this lesion had increased from 15 to 20 mm over 3 years. She was transferred to our hospital on April 8. Magnetic resonance (MR) imaging revealed a mass lesion in the pineal region that was hypointense on the T<sub>1</sub>-weighted images and hyperintense on the T<sub>2</sub>-weighted images, without enhancement by contrast medium (Fig. 3). The lesion compressed the tectal

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**Fig. 2** Preoperative computed tomography scan showing enlargement of the cystic mass in the pineal region. Obstructive hydrocephalus was improved after ventriculoperitoneal shunting via the right anterior horn.



**Fig. 4** Postoperative diffusion-weighted magnetic resonance image demonstrating the masses in the pineal and superior lateral orbital regions (arrow) as marked hyperintensity.



**Fig. 3** Preoperative magnetic resonance images showing a mass lesion hypointense on the T<sub>1</sub>-weighted (A) and hyperintense on the T<sub>2</sub>-weighted images (C), without enhancement (B), in the pineal region.

plate anteriorly, causing occlusion of the aqueduct of Sylvius. The preoperative diagnosis was pineal cyst.

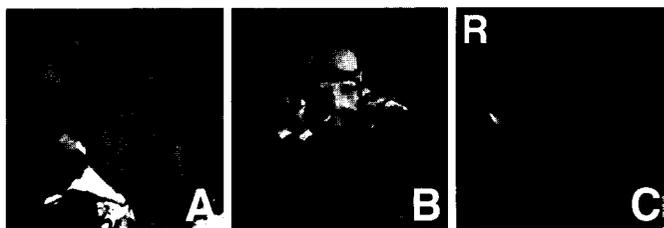
Neuroendoscopic biopsy of the cystic lesion in the pineal region and third ventriculostomy were performed through a standard coronal burr hole on April 12. A flexible fiberoptic ventriculoscope (Codman, Inc., Raynham, Mass., U.S.A.) was inserted into the anterior horn of the lateral ventricle through a peelaway sheath. The anterior wall of the lesion was recognized as a protrusion at the posterior wall of the third ventricle, which had compressed the orifice of the aqueduct of Sylvius. We first tried to perforate the anterior wall of the lesion using low-current monopolar coagulation to aspirate the contents of the cystic lesion, which were expected to be liquid. However, the wall of the lesion was very thick, so we could not perforate the wall or

remove the contents. Several specimens were taken for histological examination. Third ventriculostomy was subsequently performed.

Postoperatively, her symptoms remained unchanged, and postoperative MR imaging showed that the size of the pineal cystic lesion was also unchanged. Diffusion-weighted MR imaging demonstrated the lesions in the pineal and superior lateral orbital regions as marked hyperintensity (Fig. 4). The final neuroimaging diagnosis was multiple epidermoid cysts of the pineal region and extracranial region. The biopsy specimens were too small to enable histological diagnosis.

Two weeks after the first operation, a second endoscopic procedure was performed to obtain sufficient specimens for a definitive histological diagnosis of the cystic lesion in the pineal region and also to reduce the volume of the lesion. Endoscopically, we confirmed that the stoma was open at the tuber cinereum of the floor of the third ventricle. The white-colored solid contents of the lesion were removed to the greatest possible extent using biopsy forceps through the channel of the flexible endoscope. Histologically, the lesion was diagnosed as an epidermoid cyst.

After the second operation, her headache, somnolence, and upward gaze palsy were improved. Cerebrospinal fluid examinations revealed no abnormalities during the postoperative course. We did not perform treatment for the epidermoid cyst in the superior lateral orbital region. There has been no symptomatic or radiological evidence of regrowth or dissemination during the 5-year period following surgery. MR imaging performed 5 years after surgery demonstrated that the residual lesion in the



**Fig. 5** T<sub>1</sub>-weighted (A), T<sub>2</sub>-weighted (B), and diffusion-weighted magnetic resonance images (C) showing that the residual tumor in the pineal region had disappeared during the 5-year period following surgery.

pineal region had disappeared (Fig. 5).

### Discussion

Intracranial multiple epidermoid or dermoid cysts sometimes occur due to intrathecal seeding or trauma,<sup>5,9,10,13,17</sup> but cases with independent epidermoid cysts located in both the intracranial and extracranial regions are extremely rare. Since epidermoid cysts are thought to originate from the germinal layer and ectopic nests of epithelial cells remain in situ during closure of the neural tube,<sup>8</sup> these cysts can occur in the scalp, cranial bones, intracranial tissue, and spinal canal near the midline. Therefore, epidermoid cysts of the pineal region and the superior lateral orbital region can occur at the same time as in this case.

MR imaging usually reveals epidermoid cyst as hypointense on the T<sub>1</sub>-weighted images and hyperintense on the T<sub>2</sub>-weighted images, with no apparent contrast enhancement.<sup>6,8,14</sup> However, inclusion of water, keratin, and cholesterol in an epidermoid cyst can yield several signal intensity on MR imaging.<sup>6</sup> Diffusion-weighted imaging is useful for the diagnosis of epidermoid cysts.<sup>14</sup> In our case, diffusion-weighted imaging performed after the first operation demonstrated lesions in the pineal and superior lateral orbital regions as areas of marked hyperintensity. This finding led to the diagnosis of multiple epidermoid cysts of the pineal and extracranial regions.

For pineal epidermoid cysts, total removal is the goal of treatment. The approaches utilized include the occipital transtentorial approach,<sup>6,8,11,12,14,18</sup> infratentorial supracerebellar approach,<sup>2,8</sup> transventricular approach,<sup>16</sup> and combined supra/infratentorial-transsinus approach.<sup>19</sup> In the present case, we expected to find a pineal cyst with a thin wall and fluid content, because diffusion-weighted imaging was not performed before the first operation. A

neuroendoscopic procedure via the lateral and third ventricles<sup>4</sup> using a flexible endoscope was therefore selected. Craniotomy should be performed at the second operation after the diagnosis of epidermoid cyst. We selected endoscopic removal to the greatest possible extent in order to improve her symptoms, taking into account the minimal invasiveness of the neuroendoscopic procedure. The neuroendoscopic technique permits reliable observation of the pineal region, third ventriculostomy, and tumor biopsy in one stage. However, this procedure resulted in communication between the epidermoid cyst and the cerebrospinal fluid in the third ventricle. Leakage of cholesterol into the cerebrospinal fluid may cause chemical (aseptic) meningitis, so coagulation of epidermoid tags wherever safe is recommended to minimize the chance of reformation.<sup>3</sup> We used steroids to prevent chemical meningitis,<sup>1</sup> and fortunately meningitis did not occur. Furthermore, the residual tumor disappeared during the 5-year period following surgery. Although the communication between the residual epidermoid cyst and the cerebrospinal fluid resulting from the neuroendoscopic procedure may reduce the contents of the epidermoid cyst, we have been following the postoperative course closely.

Neuroendoscopic manipulation is minimally invasive and may be effective for treating pineal lesions. However, neuroendoscopic manipulation for a pineal epidermoid cyst is controversial because of the possibility of chemical meningitis and intrathecal seeding from the residual tumor. We suggest that careful preoperative diagnosis of the epidermoid cysts based on diffusion-weighted MR imaging is required.

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