Malignant Transformation 20 Years After Partial Removal of Intracranial Epidermoid Cyst
—Case Report—

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Abstract

A 74-year-old woman presented with malignant progression of remnant epidermoid cyst manifesting as sudden onset of right ptosis and double vision. She had right oculomotor nerve paresis. She had a history of surgery for right cerebellopontine angle epidermoid cyst 20 years previously. T1-weighted magnetic resonance (MR) imaging demonstrated a hypointense mass lesion in the right cerebellopontine angle and basal cistern, and an isointense mass in the right paraclinoid region which was strongly enhanced. Diffusion-weighted MR imaging showed hyperintense areas in the right cerebellopontine angle, ambient cistern, and basal cistern, and the paraclinoid mass as hypointense. Surgery was performed using Dolenc’s approach. Histological examination revealed that the paraclinoid tumor adjacent to the epidermoid tumor remnant was malignant transformation of epidermoid cyst into squamous cell carcinoma. She was treated with 46 Gy linac radiotherapy. She has been without tumor recurrence for 17 months. Malignant change of epidermoid cyst is extremely rare, but rapid progress of the symptoms suggests malignant transformation. MR imaging with gadolinium is useful for diagnosis.

Key words: epidermoid cyst, malignant change, squamous cell carcinoma, diffusion-weighted magnetic resonance imaging, oculomotor nerve paresis
**Introduction**

Intracranial epidermoid cysts are rare and account for 0.2–1.8% of all intracranial tumors. The target of surgery is complete removal of the epidermoid cyst, but it is better to leave minute capsule remnants than risk a surgical catastrophe. Epidermoid cyst is lined with benign keratinizing squamous epithelium and malignant transformation of remnant epidermoid cyst long after the initial surgery is extremely rare. Here we describe a case of an epidermoid cyst in the cerebellopontine (CP) angle and basal cistern which underwent malignant transformation 20 years after removal.

**Case Report**

A 74-year-old woman had a history of right CP angle epidermoid cyst manifesting as headache and dizziness, and had undergone surgery at a local hospital 20 years previously. Since then, she had suffered no neurological deficit, but was not followed up regularly. She suffered sudden onset of right ptosis and double vision. She visited a local clinic and magnetic resonance (MR) imaging revealed an enhanced right paracclinoid mass lesion.

On admission to our hospital, she had right oculomotor nerve paresis, but the other cranial nerves were intact. T1-weighted MR imaging demonstrated a hypointense mass lesion in the right CP angle and an isointense mass in the right paracclinoid region (Fig. 1A). The right paracclinoid mass lesion was strongly enhanced after gadolinium-diethylenetriaminepenta-acetic acid administration (Fig. 1B). Diffusion-weighted MR imaging showed hyperintense areas in the right CP angle, ambient cistern, and basal cistern. The paracclinoid mass appeared as hypointense adjacent to the hyperintense area in the basal cistern (Fig. 1C). We considered that the paracclinoid mass was the cause of her right oculomotor nerve paresis.

Right frontotemporal craniotomy and right anterior clinoidectomy via Dolenc’s extradural approach were employed to access the paracclinoid lesion. Slightly yellowish pearly tumor was observed in the basal and prechiasmatic cisterns (Fig. 2A). Adhesion to the surrounding brain tissue was not strong. The flaky pearly tumor was subtotally removed. Pinkish paracclinoid mass lesion adjacent to the basal pearly tumor was exposed with a relatively clear border between the pinkish tumor and pearly tumor. The elastic hard pinkish tumor was totally removed (Fig. 2B). Histological examination revealed that the pearly tumor was consistent with benign epidermoid cyst (Fig. 3D), and the adjacent paracclinoid tumor consisted of a mixture of degenerated squamous epithelium and high pleomorphic cellularity with mitotic activity (Fig. 3A, B). Immunoreactivity for cytokeratin 5/6 was strong (Fig. 3C). The diagnosis of the paracclinoid lesion was malignant transformation of benign epidermoid cyst to squamous cell carcinoma.

The postoperative course was uneventful. She was treated with 46 Gy linac radiotherapy to the basal cistern. MR imaging performed 17 months after the operation showed...
Malignant transformation of epidermoid cyst into carcinoma is quite rare. Most previous cases of these carcinomas were discovered within benign epidermoid cysts at the first surgery or autopsy. Malignant transformation from remnant epidermoid cyst long after the initial surgery is extremely rare with only 8 previous cases (Table 1). The patients were aged from 36 to 74 years (mean 54 years), and 7 of the 9 including our case were female. The interval from first operation to malignant transformation ranged from 2 to 33 years (mean 15.5 years). Epidermoid cysts undergoing malignant transformation after a short period have been reported. However, squamous cell carcinomas were probably already present in most cases.

Rapid progression of symptoms and signs is the most important clinical indication of malignant transformation of epidermoid cysts. Typical imaging findings are rapid growth of the focal enhanced part within the mass on computed tomography or MR imaging. Benign epidermoid cyst appears as hyperintense on diffusion-weighted MR imaging and is useful for diagnosis and postoperative follow up. The malignant part of the epidermoid cyst appears as hypointense on diffusion-weighted MR imaging. Diffusion-weighted MR imaging of our case also demonstrated a hypointense area adjacent to the hyperintense benign epidermoid area.

Differential diagnosis between malignant transformation of epidermoid cyst and foreign body giant cell reaction is difficult. Foreign body giant cell reaction also appears as focal enhanced mass adjacent to epidermoid cyst. Therefore, histological diagnosis is necessary before adjuvant therapy. The exact mechanisms of malignant change of epidermoid cyst remain unclear. Chronic inflammatory response to repeated cyst rupture and subtotal resection of the cyst wall may cause the malignant transformation. Radiation therapy after surgical resection of malignant epidermoid cyst seems to be effective. Four of the eight patients received radiotherapy with mean follow up of 31.5 months (Table 1). On the other hand, the four patients not treated with irradiation had mean follow up of only 1.7 months. Radiation therapy must be considered after surgery. Recently, gamma knife radiosurgery for adjuvant therapy was also reported as useful.

The present case of malignant transformation of benign epidermoid cyst illustrates that rapid progress of the symptom suggests malignant degeneration of the epidermoid cyst. MR imaging with contrast medium is useful for diagnosis. Physicians should respond to rapid symptom changes in any patient who previously underwent surgery for benign epidermoid cyst, and arrange for MR imaging with gadolinium. Surgical resection and adjuvant radiation therapy are highly recommended.

References

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Table 1 Cases of malignant transformation of epidermoid cyst

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author (Year)</th>
<th>Age (yrs)/Sex</th>
<th>Tumor location</th>
<th>Treatment</th>
<th>Interval (yrs)</th>
<th>Postoperative course</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Fox and South (1965)</td>
<td>43/M</td>
<td>temporal</td>
<td>S</td>
<td>7</td>
<td>died 1 mo</td>
</tr>
<tr>
<td>2</td>
<td>Goldman and Gandy (1987)</td>
<td>59/F</td>
<td>intraventricular</td>
<td>S, Rx</td>
<td>33</td>
<td>alive 36 mos</td>
</tr>
<tr>
<td>3</td>
<td>Abramson et al. (1989)</td>
<td>36/M</td>
<td>CP angle</td>
<td>S</td>
<td>2</td>
<td>ND</td>
</tr>
<tr>
<td>4</td>
<td>Tognetti et al. (1991)</td>
<td>67/F</td>
<td>frontotemporal</td>
<td>S</td>
<td>31</td>
<td>died 1 mo</td>
</tr>
<tr>
<td>5</td>
<td>Murase et al. (1999)</td>
<td>50/F</td>
<td>CP angle</td>
<td>S, Ch, Rx</td>
<td>10</td>
<td>alive 60 mos</td>
</tr>
<tr>
<td>6</td>
<td>Asahi et al. (2001)</td>
<td>55/F</td>
<td>CP angle</td>
<td>S</td>
<td>13</td>
<td>died 3 mos</td>
</tr>
<tr>
<td>7</td>
<td>Nawashiro et al. (2001)</td>
<td>46/F</td>
<td>temporal</td>
<td>S</td>
<td>ND</td>
<td>ND</td>
</tr>
<tr>
<td>8</td>
<td>Tamura et al. (2006)</td>
<td>50/F</td>
<td>CP angle</td>
<td>S, Rx</td>
<td>8</td>
<td>alive 13 mos</td>
</tr>
<tr>
<td>9</td>
<td>Present case</td>
<td>74/F</td>
<td>CP angle</td>
<td>S, Rx</td>
<td>20</td>
<td>alive 17 mos</td>
</tr>
</tbody>
</table>

Ch: chemotherapy, CP: cerebello-pontine, ND: not described, Rx: radiation therapy, Rx*: gamma knife, S: surgery.
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