

# MR Imaging of a Cerebello-Pontine Angle Epidermoid Cyst with a Malignant Transformation: Case Report<sup>1</sup>

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The malignant transformation of an epidermoid cyst can be discovered at the same time as a pre-existing epidermoid cyst, or during a follow-up examination after an incomplete excision. We describe a rare case of malignant transformation of a cerebello-pontine angle epidermoid cyst which extended into the middle cerebellar peduncle.

**Index words :** Brain neoplasms

Brain, MR

Neoplasms, squamous cells

Epidermoid cysts account for 0.2 - 1.8% of all intracranial tumors and are histologically benign (1). A case of the transformation of an epidermoid cyst into a malignant squamous cell carcinoma has seldom been reported (2 - 7). The malignant transformation may have occurred from pre-existing epidermoid cysts or after the incomplete excision of an epidermoid cyst (5). The treatment of the malignant transformation of an epidermoid cyst varies from surgery or a combination of treatments such as radiotherapy and chemotherapy, however, patient prognosis is poor. The malignant transformation of an epidermoid cyst has characteristic MR imaging findings and a clinical course. We report the MR imaging findings of a malignant transformation of a cerebello-pontine angle epidermoid cyst.

## Case Report

A 69-year-old woman visited the neurosurgery department at the College of Medicine Yeungnam University

Hospital due to long standing dizziness and the recent development of right facial hypoesthesia. The MR imaging revealed a mass located between the right middle cerebellar peduncle and the cerebello-pontine angle. This mass appeared hyperintense on T2-weighted images, and hypointense on T1-weighted images (Fig. 1A, B). A contrast study revealed a rim and nodular enhancement in the middle cerebellar peduncular portion; however, we could not find the contrast enhancement in the cerebello-pontine angle mass (Fig. 1C). A diffusion-weighted image revealed that the mass produced a low signal intensity in middle cerebellar peduncle and a high signal intensity in cerebello-pontine angle (Fig. 1D). During the operation, an extra-axial mass with pearly desquamated contents was found at the right cerebello-pontine angle. Also, we found the well encapsulated extra-axial mass, which was tightly adhered to the middle cerebellar fossa; hence, total removal was impossible.

The histopathological findings of the resected specimen obtained from cerebello-pontine angle showed keratinous material which was characteristic of an epidermoid cyst. The specimen obtained from the middle cerebellar peduncle demonstrated a well differentiated squamous carcinoma with cellular pleomorphism and a mitotic feature.

A follow-up MRI obtained 10 months after surgery re-

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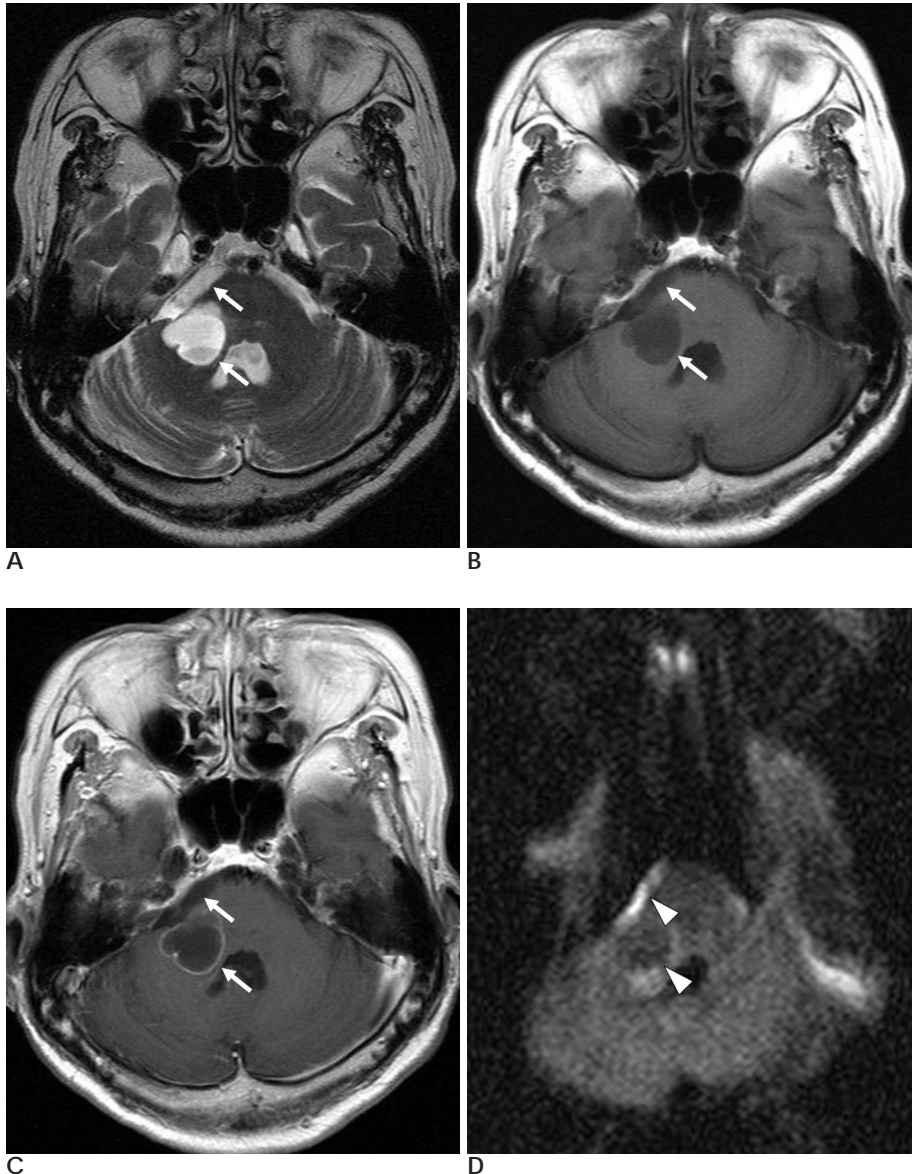
vealed a peripheral enhancing mass with a central necrotic portion in the right cerebello-pontine angle. The patient was treated with adjuvant radiotherapy to eliminate any residual tumor material.

### Discussion

The malignant transformation of a benign cyst such as an epidermoid cyst is rare. It is however the third most common cause of intracranial squamous cell carcinoma following metastasis, as a direct extension from the cranial base (8). The exact mechanism of the malignant transformation of an epidermoid cyst is still unknown; however, chronic inflammation after incomplete surgery or repeated rupture and in situ squamous metaplasia of an epidermoid cyst are suggested as possible causes (8).

Five types of malignant transformations from a benign intracranial cyst are suggested: initial malignant transformation of an epidermoid cyst, malignant transformation from a remnant epidermoid cyst, malignant transformation with leptomeningeal carcinomatosis, squamous cell carcinoma arising from other benign cysts, and other malignancies arising from a benign cyst (8). Above all, the most frequent type of malignant transformation is the initial malignant transformation of an epidermoid cyst. That is, the carcinoma is discovered during the first resection of an epidermoid cyst or during an autopsy (7, 8).

Clinically speaking, if a patient is rapidly disoriented and does not quickly recover after the surgery for the removal of a benign cyst or rapid recurrence and the occurrence of leptomeningeal carcinomatosis, the malignant transformation of an epidermoid cyst must be con-



**Fig. 1. A-D.** The mass is located from the right middle cerebellar peduncle to the cerebello-pontine angle (arrows). The MR images revealed that the identified mass produces a high signal intensity on a T2-weighted image (A) and a low signal intensity on a T1-weighted image (B). The mass in the right middle cerebellar peduncle shows rim and nodular enhancement. In addition, the cerebello-pontine angle mass does not demonstrate contrast enhancement (C). The diffusion weighted image shows high signal intensity in the cerebello-pontine angle and low signal intensity in middle cerebellar peduncle (arrowheads) (D).

sidered (7, 8).

A computed tomography (CT) scan shows a hypodense mass and rim enhancement in the cerebello-pontine angle. Enhancement is also noted in pathologically benign tumors because of peritumoral granulation (9). The primary signs of the malignant transformation of an epidermoid cyst is a focal enhancement at the tumor after administering the contrast medium (2 - 9). Edema and leptomeningeal metastasis may be found during the first examination, as well as the follow up. For a diffusion-weighted image, the signal intensity of an epidermoid cyst is higher than that of the cerebrospinal fluid; however, the malignant transformed portion of the mass shows a low signal intensity, which corresponds to central necrosis (4). A differential diagnosis must be made in case of a foreign body giant cell reaction, because a ruptured epidermoid cyst causes inflammation in the adjacent tissue and can show nodular enhancement similar to the malignant transformation of an epidermoid cyst (10).

According to the systemic review of articles pertaining to the malignant transformation of an intracranial benign epithelial cyst, the prognosis is poor. Thirty nine out of the 52 patients died with most patients dying within 12 months after the symptom onset or the diagnosis (8). Complete excision is the treatment of choice (7, 8). But, complete excision is limited to when the capsular adhered to perforators, cranial nerve, and brain stem are found at surgery (5). The radiation therapy, chemotherapy or combination therapy has been found to extend patient survival (5 - 7).

In conclusion, the malignant transformation of an epidermoid cyst must be considered if the rim or nodular enhancement on the epidermoid cyst is demonstrated at first or during follow up after surgery. The diffusion-

weighted image is also helpful in finding an epidermoid cyst and to detect the malignant transformation of an epidermoid cyst. In addition, even if the epidermoid cyst was completely removed, careful clinical and radiological observation must be made to determine the early detection of the malignant transformation of an epidermoid cyst.

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