

# Malignant transformation of an epidermoid cyst in the temporal and prepontine region: Report of a case and differential diagnosis

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**Abstract.** Malignant transformation of an epidermoid cyst is rare. The current report presents a case of a 55-year-old female patient with squamous cell carcinoma arising from a benign epidermoid cyst in the left temporal region and prepontine area. She had undergone subtotal resection of an epidermoid cyst 7 months previously. Preoperative imaging findings included a focal enhancing area adjacent to the lesion. Postoperative computed tomography demonstrated an increase in the size of the enhancing area. The patient underwent removal of the lesion and postoperative histological examination revealed a squamous cell carcinoma possibly arising from the epidermoid cyst. Accurate diagnosis of malignant transformation prior to operation is difficult; however, the possibility of an intracranial epidermoid cyst must be considered if a focally enhancing area is visible. Postoperative histological examinations may be used to determine a definite diagnosis, and accurate diagnosis is important for planning a rational therapeutic strategy.

## Introduction

Epidermoid cysts are benign intracranial tumors that are predominantly located in the cerebellopontine angle, accounting for ~1% of intracranial tumors (1). The cysts are lined with benign keratinizing squamous epithelium and contain laminated keratin. Typical magnetic resonance imaging (MRI) features include lesions with signal intensity slightly greater than that of the cerebrospinal fluid on T1-weighted imaging and high density on T2-weighted imaging without contrast enhancement (2,3). The presence of contrast enhancement within or adjacent to an epidermoid cyst is rare, but may indicate various conditions including giant cell

reaction, coexistence of different histological types and malignant transformation of the epidermoid cyst (4-6).

The current study presents a case of pathologically confirmed epidermoid cyst with a focal enhancing area adjacent to the lesion observed on MRI. The diagnostic procedure and differential diagnosis are discussed. Written informed consent was obtained from the patient's family.

## Case report

In November 2013, a 55-year-old woman visited the Neurosurgical Clinic at the Renji Hospital, Shanghai Jiaotong University School of Medicine (Shanghai, China) due to intermittent headaches and progressive right limb weakness in both limbs. Physical examination upon admission revealed that the muscle weakness in the right limb was grade 3, according to the modified Medical Research Council grading scale (7). Computed tomography (CT; Aquilion™ Vision; Toshiba, Tokyo, Japan) of the head demonstrated a sharply defined, low-density lesion occupying the left temporal region and prepontine area (Fig. 1). MRI of the head (Signa Excite System 3.0T; GE Healthcare Bio-Sciences, Pittsburgh, PA, USA) revealed a large cystic mass in the left temporal region and prepontine area, which was hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging, with focal enhancement adjacent to the thalamus region, a 5-mm shift of midline structure from right to left, and brainstem compression (Fig. 1). The mass was approached via a left modified trans-pterional approach. The focally enhanced part of the mass was not resected due to its close association with the basal ganglia and its possible pathological nature of a giant cell reaction. The lesion appeared to have the typical pearly-white gross appearance of an epidermoid cyst. The resected tissues were fixed in 10% formalin, paraffin-embedded and cut into 5-mm sections for immunohistochemical analysis. Hematoxylin-eosin (Solarbio Co., Ltd., Beijing, China) staining revealed that the cyst was lined by keratinizing squamous epithelium and contained lamellated keratinous debris; no malignant change was suspected from the pathological findings of the resected specimen (Fig. 2A). A postoperative CT scan of the head revealed disappearance of the cyst, and remnants of the enhancing part of the lesion. The patient's symptoms were markedly improved following the surgery (Fig. 1).

Intermittent CT of the head subsequent to the surgery demonstrated that the size of enhancing part had gradually

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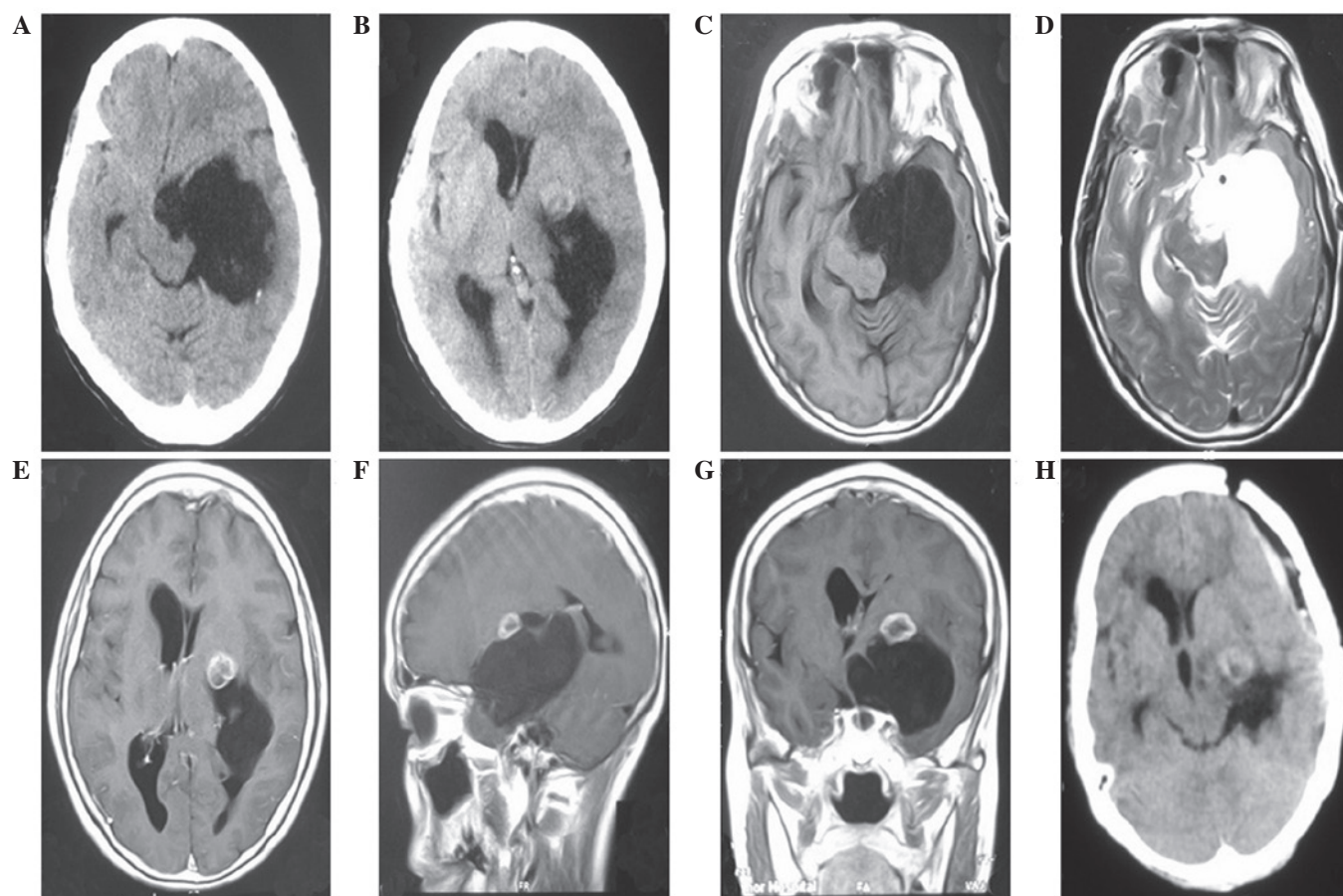


Figure 1. Pre- and post-operative imaging findings. (A and B) Preoperative CT of the head demonstrated a sharply defined low-density lesion occupying the left temporal region and prepontine area. (C-G) Sagittal MRI of head revealed a large cystic mass of left temporal region and prepontine area, which was (C) hypointense on T1WI and (D) hyperintense on T2WI, with focal enhancement adjacent to the thalamus region, a 5-mm shift of midline structure from right to left, and brainstem compression. (E) T1WI sagittal MRI with contrast, (F) T1WI axial MRI with contrast and (G) T1WI coronal MRI with contrast reveals a focal enhancing area adjacent to the lesion. (H) Postoperative CT of head showed disappearance of cyst and remnants of the enhancing part of the lesion. CT, computed tomography; MRI, magnetic resonance imaging; T1WI, T1-weighted imaging; T2WI, T2-weighted imaging.

increased in comparison with that on the immediate post-operative CT (Fig. 2B-D). However, the patient refused reoperation until paralysis of the right limb occurred ~7 months later. Following re-admission, MRI of the head revealed that the lesion occupied the left temporal lobe, appeared hypointense on T1-weighted images and had mixed intensity on T2-weighted images, with edge enhancement and hydrocephalus (Fig. 3A-E). The lesion was resected via the aforementioned approach. Postoperative histological examination of the lesion indicated squamous cell carcinoma (Fig. 3F). A metastatic work-up was conducted subsequently to look for a primary focus elsewhere, but no metastatic lesions were observed in other organs of the body.

The lesion was clinically diagnosed as a malignant transformation of an epidermoid cyst. Adjuvant radiotherapy was recommended. However, the patient refused adjunctive therapies for personal reasons. The patient succumbed to pneumonia at ~6 months following the second surgery.

## Discussion

According to a systemic review conducted by Hamlat *et al* (1) and the current findings, present case is classified as the first type of malignant transformation: Initial malignant

Table I. Criteria for diagnosis of primary squamous cell carcinoma.

No.	Description
1	Tumor restricted to the intracranial intradural compartment
2	No invasion of or extension beyond the dura or cranial bones or through cranial orifices
3	No communication with the middle ear, air sinuses or sella turcica
4	No evidence of a nasopharyngeal tumor
5	Presence of benign squamous epithelium within the main tumor mass
6	No evidence of a primary tumor elsewhere

transformation of an epidermoid cyst. Malignant transformation of an epidermoid cyst is a well-documented but rare occurrence (2,6). The rapid progression of signs and symptoms is the most important clinical indication of malignant transformation of epidermoid cysts (8). The exact mechanism of malignant changes of epidermoid cysts remain unclear.



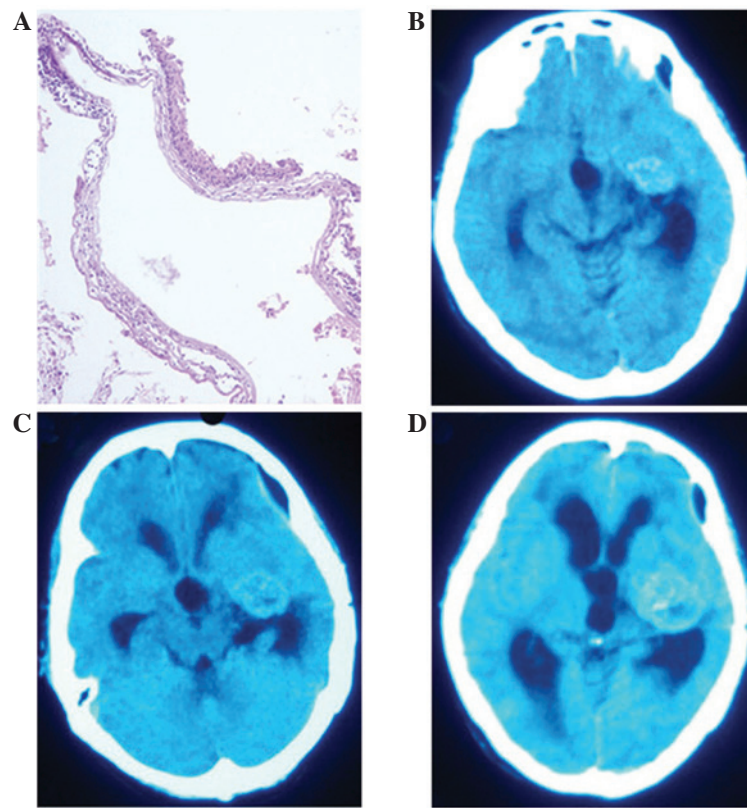


Figure 2. Repeated CT of the head and postoperative histopathological examinations. (A) The cyst wall of specimen showed multilayered keratin debris with enlarged pleomorphic nuclei. (B-D) Consecutive postoperative CT of head demonstrated that the enhancing part of the lesion had gradually increased in comparison with postoperative CT. CT, computed tomography.

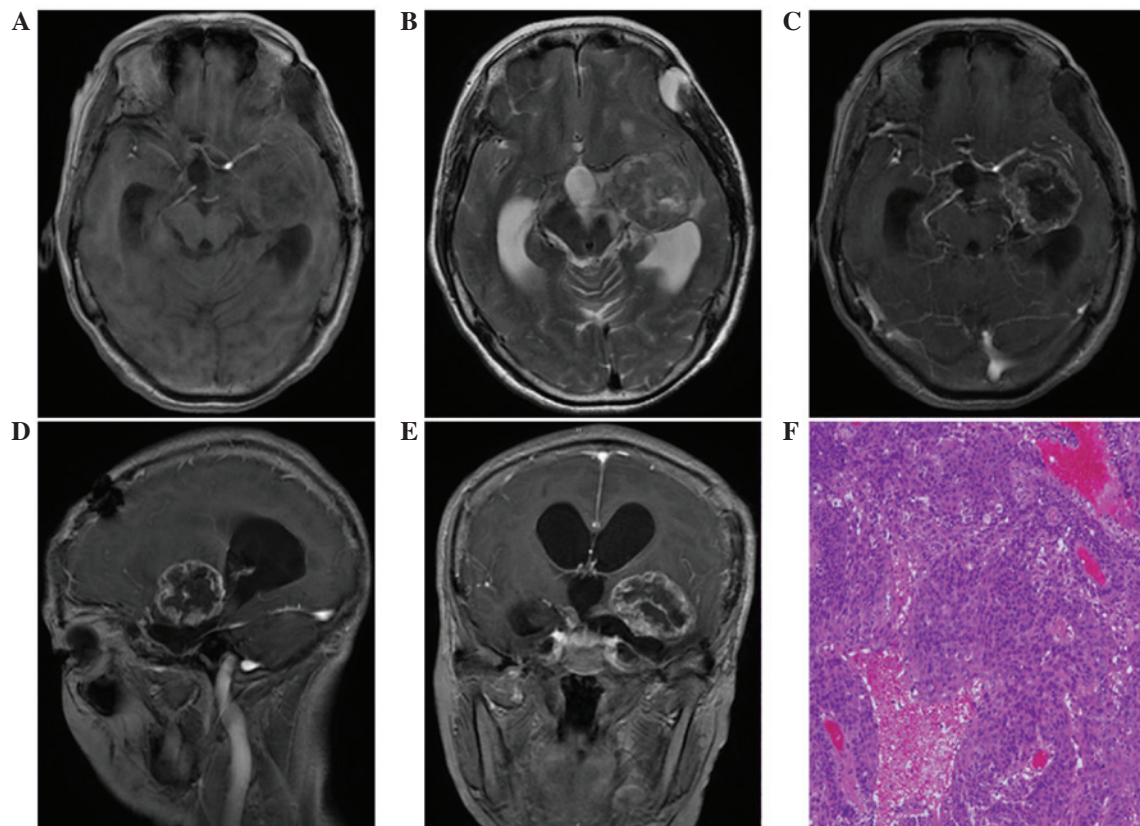


Figure 3. Imaging findings prior to second operation. (A-E) Sagittal MRI of the head showed the lesion occupying the left temporal lobe, which was (A) hypointense on T1WI and (B) hyperintense on T2WI, with edge enhancement and hydrocephalus. (C) T1WI sagittal MRI with contrast, (D) T1WI axial MRI with contrast and (E) T1WI coronal MRI with contrast reveals an enhancing lesion in the left temporal region. (F) Irregular atypical cells and keratinization (hematoxylin and eosin staining; magnification, x100). MRI, magnetic resonance imaging; T1WI, T1-weighted imaging; T2WI, T2-weighted imaging.

Chronic inflammatory response to repeated cyst rupture and subtotal resection of the cyst wall may result in malignant transformation (9).

Diagnosis of malignant transformation of an epidermoid cyst is difficult, particularly when solely dependent on imaging findings. Characteristic imaging findings of malignant transformation of an epidermoid cyst include a focal enhancing region within the mass on CT or MRI (2). In addition to typical imaging findings, diffusion-weighted imaging may help to distinguish the nodular enhancing area (2,10). Previous studies have reported that malignant parts of an epidermoid cyst exhibit a low signal, whereas benign regions exhibit a very high signal. Possible explanations include central necrosis in the mass lesion and the T2-shine-through effect (11). Accurate diagnosis of malignant transformation of an epidermoid cyst primarily depends on postoperative histopathological examination. Garcia *et al* (12) and Hamlat *et al* (1) summarized the criteria for its histological diagnosis (summarized in Table I). Hamlat *et al* (1) conducted a systematic review and found that 70.3% of patients (52 of 74) fulfilled Garcia's criteria (1).

Differentiation between malignant transformation of epidermoid cyst, giant cell reaction and coexistence of different histological types on imaging findings is challenging (2). Leakage of cyst content, including cholesterol, keratin and cellular debris, into the subarachnoid space upon spontaneous rupture of the epidermoid cyst may result in chemical meningitis. In such a situation, leptomeningeal enhancement may be observed on post-contrast MRI (2). Foreign giant cell reaction, an inflammatory giant cell reaction of the ruptured epidermoid cyst, may also occur in the brain parenchyma (2). Moran *et al* (4) suggested that the epidermoid cyst in their patient may have ruptured spontaneously into the adjacent brain parenchyma, leading to an intense local inflammatory response with the presence of multinucleated giant cells (4). In such cases, MRI may show an enhanced region with associated vasogenic edema. As for the coexistence of different histological types, to the best of our knowledge, there has only been one case reporting the adjacent nodular enhancing region and epidermoid cyst (5).

The prognosis of malignant transformation of an epidermoid cyst is poor (6). Based on a previous review, the mortality rate was as high as 75% (39 of 52 patients) (2). Surgery followed by radiotherapy seems to be the optimal therapeutic modality (13). However, in the present case, the patient declined our recommended treatment for personal reasons and succumbed to disease within 12 months of the surgery.

The current study presents a rare case of malignant transformation of an epidermoid cyst in the temporal and prepontine region. Providing an accurate diagnosis of this type of tumor prior to surgery is challenging; however, the possibility of malignant transformation should be considered if the focal enhancing area is visible. Comprehensive therapy is recommended for patients with a definite diagnosis.

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