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*Case Report*

# Surgical treatment and radiotherapy of epidermoid cyst with malignant transformation in cerebellopontine angle

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**Abstract:** We report surgical treatment and radiotherapy of an extremely rare case of malignant epidermoid cyst located in cerebellopontine angle. MRI and CT demonstrated the lesion with partial enhancement and calcification. During operation, we found the tumor attached tightly to surrounding tissue. Finally we achieved near total resection of it. Histopathology confirmed the diagnosis of epidermoid cyst with malignant transformation. With adjuvant radiotherapy, the patient underwent excellent recovery, and follow-up MRI demonstrated no obvious residue or recurrence of the tumor. Malignant epidermoid cyst can be diagnosed radiologically in combination with clinical presentation. Maximal removal plus adjuvant radiotherapy is the treatment of choice, although the general prognosis of it is poor.

**Keywords:** Intracranial epidermoid cyst (IEC), malignant epidermoid cyst (MEC), surgical treatment, intensity-modulated radiotherapy (IMRT)

## Introduction

Generally, intracranial epidermoid cysts (IECs) are benign and slow growing lesions accounting for about 1% of all intracranial tumors [1-3]. Cerebellopontine angle (CPA) is one of the most common sites of IEC which constitute approximately 7% of tumors in this region [2]. Malignant transformation of IEC is extremely rare, and little is known about its clinical, radiological features and prognosis. We present a case of malignant epidermoid cyst (MEC) in CPA who have acquired good recovery after successful surgery and radiotherapy, relevant literature is also reviewed. To our knowledge, since we performed near-total resection of the tumour, we happen to provide the very experience of intensity-modulated radiotherapy (IMRT) of this rare clinical entity.

## Case report

This 42-year-old male suffered from hearing decrease since 20 years ago and never accepted radiological exams. Until recently, he found it's difficult to write normally or to walk stably, then he underwent head magnetic resonance imaging (MRI) which demonstrated a huge tumour located in right CPA (**Figure 1A**, **1B**). The diagnosis at admission was "neurinoma with haemorrhage or cystic change". Computerized tomography (CT) demonstrated no obvious enlargement of internal auditory canal (IAC) or bone damage, but spotty hyperintensive calcification is present (**Figure 1C**). Neurological examination mainly showed right pyramid sign and right hearing loss. Intraoperatively, part of the tumour was like typical IEC which is pearly and easy to remove, while the other part was tenacious in texture and rich in blood supply, adhering very tightly to surrounding brain structures. It took a very gentle and skilled maneuver to detach the tumour by pieces (**Figure 2A**). Tumour within IAC was also pulled out carefully (**Figure 2B**). Finally we achieved near-total removal of the tumour with little potential remnant capsule left on cranial nerves or arteries

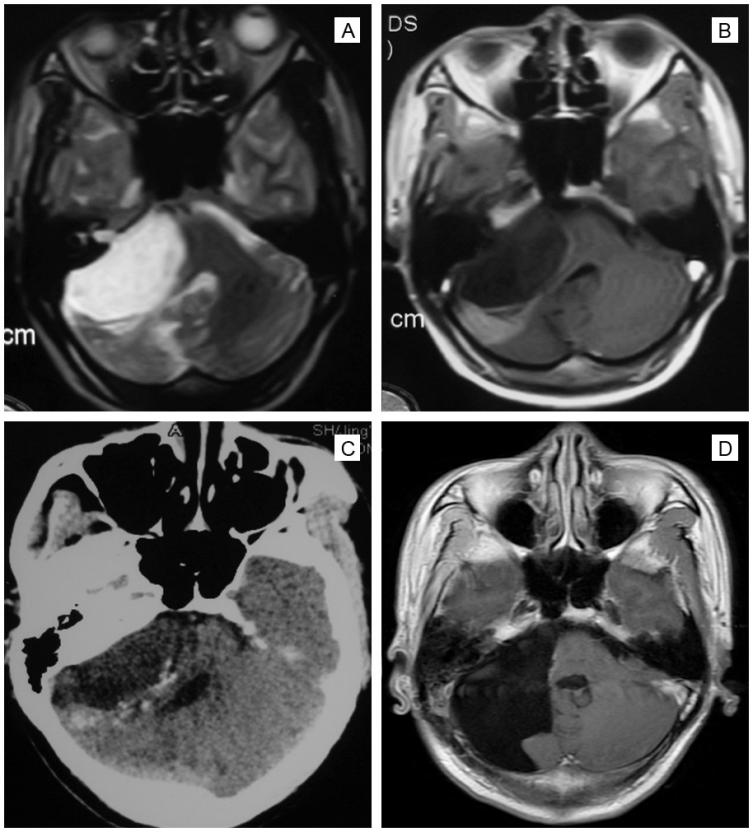


**Figure 1.** Pre- and post-operative MRIs. T2WI (A) before operation showed a hyperintensive tumour located in the CPA. Gd-DTPA enhanced MRI (B) presented the tumour with partial enhancement. CT scan (C) before operation demonstrated hyperintensity in the tumour, indicating calcification. Gd-DTPA enhanced MRI (D) 6 months after operation demonstrated no visible tumour residue.

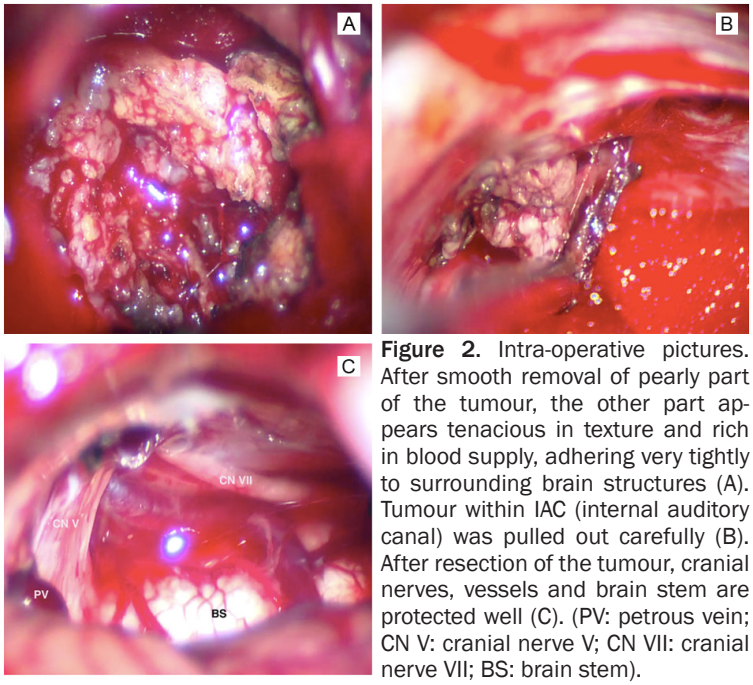


**Figure 2.** Intra-operative pictures. After smooth removal of pearly part of the tumour, the other part appears tenacious in texture and rich in blood supply, adhering very tightly to surrounding brain structures (A). Tumour within IAC (internal auditory canal) was pulled out carefully (B). After resection of the tumour, cranial nerves, vessels and brain stem are protected well (C). (PV: petrous vein; CN V: cranial nerve V; CN VII: cranial nerve VII; BS: brain stem).

(**Figure 2C**). After operation there's no further neurological deterioration. Histopathology confirmed the diagnosis of MEC (**Figure 3**). One month after operation, since there is no visible tumor residue, our radiotherapist recommended IMRT rather than Gama-knife (which is mostly chosen in literature) with a dose of 36 gray (Gy) in 12 fractions to the patient. He recovered excellently at 6-month follow-up with facial symmetry, stable gait as well as normal muscular strength, and enhanced MRI (**Figure 1D**) showed absence of tumour and no signs of recurrence.

## Discussion

IEC in CPA usually presents with symptoms of cranial nerve irritation such as trigeminal neuralgia. However, patients with MEC usually suffered from more aggressive symptoms like hearing deficit and pyramidal defect, simulating huge acoustic neurinoma [4]. Rapid deterioration of symptoms and absence of recovery after surgery may imply MEC [3, 5]. There is evidence supporting that IEC has the tendency to malignant transformation even without surgical interference [6]. Although without imaging evidence, we agree that because for our case the patient had a history of hearing decrease as long as 20 years, which indicates that the tumor started benign but then turned to malignancy, since malignant tumor won't have such a long disease history.

Radiologically, enhancement of a epidermoid cyst can be a marker of malignant transformation [7]. Typically, CPA epidermoid cyst is easily diagnosed in MRI [8]. When

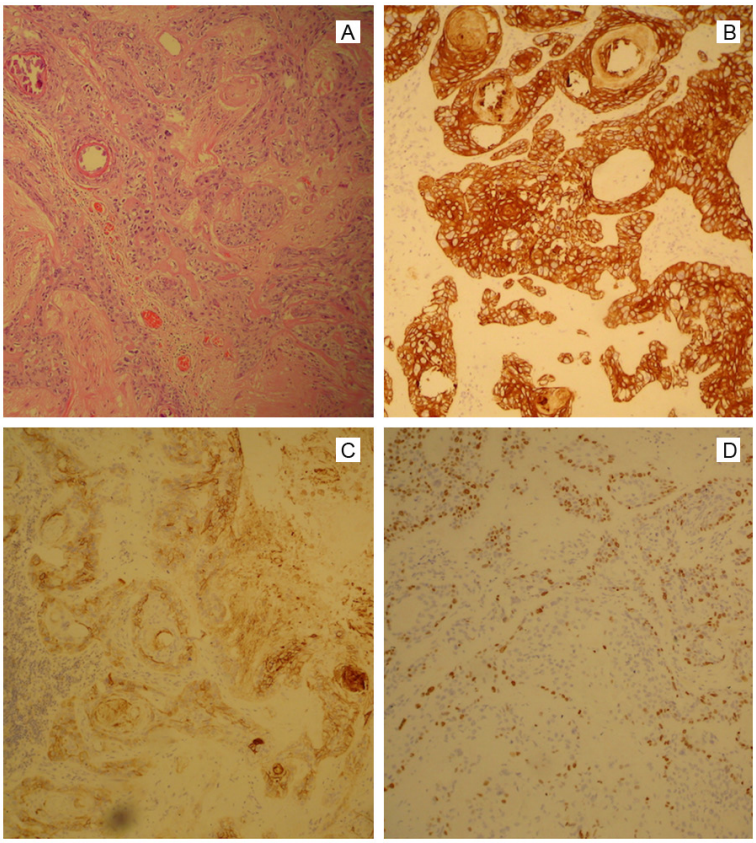


**Figure 3.** Histopathological findings. Cells of squamous carcinoma proliferate along leptomeninges, with nucleolus heteromorphism and high mitotic activity. Infiltration of inflammatory cells and calcification can be found in some specific areas (Hematoxylin-Eosin, A, ×100). Immunohistochemistry test showed that cytokeratin (B, ×100) and epithelial membrane antigen (C, ×100) are positive. MIB-1 index (D, ×100) is approximately 20%.

epidermoid cyst is radiologically suspected and there happens to be enhanced part of the lesion, then a diagnosis of MEC should be highly suspected [1, 3, 7]. Such enhanced part usually indicates malignant transformation of the tumor. In addition, cerebellar and pons edema around the tumor is deemed as another indicator for malignant transformation [7]. MEC should be differentiated from ruptured dermoid cyst. The latter should present high signal component within the tumor and scattered small "spots" in subarachnoid space or ventricular system on T1 weighted image (T1WI), but the tumor itself shouldn't be enhanced. For MEC, scattered high signal components beyond the tumor on T1WI usually can not be seen, and the enhanced part of the tumor tend to be isointense on T1WI [9].

Histopathologically, tumor cells of MEC are generally well demarcated and irregular in size with enlarged pleomorphic nuclei and increased mitotic activity. The carcinoma cells either circumscribe or infiltrate surrounding tissue [3, 7]. Immunohistochemically, the tumor cells may react with antibodies to cytokeratin (CK) and epithelial membrane antigen (EMA) [3]. Furthermore, one should notice that If histopathology or surgical removal miss the enhanced part of the tumor, then a wrong diagnosis of benign tumor can possibly be made [4].

Resective surgery, either for diagnosis confirmation or maximal removal, is the primary treatment of MEC, especially when the tumor is huge in volume. However, generally total removal is difficult to achieve in most cases for tumours' tight adherence to surrounding structures [7, 9]. We ascribe our success in maximal resection to that the malignant component of our case attached mainly to cerebellum rather than the brain stem, making it possible to perform expanded resection. IEC is usually not a candidate for radiotherapy. However, although still in controversy, benefits of radiosurgical treatment to MEC has been highlighted [4, 9-12]. Radiosurgery such as Gamma-knife surgery is mostly chosen because in most cases total or near-total removal is impossible and small residue of tumour is left. Since we manage to remove the tumour to the largest extent, we happen to provide the very experience of IMRT to this clinical entity. Usually radiotherapy can only provide temporary shrinkage of the tumor [7], but there is literature supporting that with adjuvant radiotherapy after surgery, the survival of patients would possibly extend [6]. We believe that maximal safe resection of the tumor plus radiotherapy after surgery should be treatment of choice. Regrettably, even with such sufficient treatment, the general prognosis of MEC is poor. According to a systematic review by Hamla et al, the overall median survival of MEC is 9 months [3].

## Conclusion

MEC can be diagnosed by MRI, clinical presentation and finally histopathology. Although according to literature, general prognosis of this clinical entity is poor, we believe that maximal resective surgery with uttermost protection of intracranial structures plus adjuvant radiotherapy can yield best outcome.

## Disclosure of conflict of interest

None.

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