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Case Report

Cystic Meningioma in the Cerebellopontine Angle Mimicking Vestibular Schwannoma: A Case Report

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ABSTRACT

Cystic meningioma in the cerebellopontine angle (CPA) is an extremely uncommon disease. It is often misdiagnosed as other diseases. Its clinical features, surgical strategies and prognosis are not clearly understood. We reported a case of cystic meningioma in the CPA with tumor invasion into the internal auditory canal (IAC). Based on the typical preoperative symptoms, signs, hearing tests, and enhanced magnetic resonance imaging (MRI), the 36-year-old female patient was diagnosed with vestibular schwannoma and underwent surgical resection. Postoperative pathology revealed that the tumor was meningioma. It was totally removed without any new neurological dysfunction, and no recurrence was observed in the follow-up within 24 months. Cystic meningioma in the CPA is considered to have a high pathological grade and recurrence rate. Considering this situation, total intraoperative resection, including the enhanced wall of the tumor and postoperative follow-up may be critical.

Introduction

Meningioma, originating from the arachnoid cells of the pia mater, is a common intracranial neoplasm, which represents 13-18% of all intracranial tumors [1]. At present, surgical removal of the tumor is the standard treatment. However, cystic meningioma is an uncommon disease and mostly occurs in the supratentorial region [2]. There are few reports focused on cystic meningioma located in the CPA, a sub tentorial area. So far, we have little knowledge about the clinical features, treatment strategies and prognosis of cystic meningioma in the CPA. We reported a case of CPA cystic meningioma mimicking the vestibular schwannoma.

Case Report

A 36-year-old woman suffered from intermittent dizziness and headache for 1 month and had unsteady gait for 3 days. No obvious abnormalities were found in routine laboratory tests. The preoperative hearing test suggested sensorineural hearing loss at the right ear, especially in the high-frequency band. Head computed tomography (CT) and Gd-enhanced MRI showed a cystic lesion of 5.2×3.6×3.2cm in the right CPA, with an unevenly reinforced nodule entering the internal auditory canal (Figure 1). In addition, preoperative cranial computed tomography angiography (CTA) revealed no hypervascular lesions. Based on the patient's typical symptoms and signs, as well as the results of the audiogram and craniocerebral MRI, the preliminary diagnosis was vestibular schwannoma.

During the surgery, the solid mass had abundant blood supply. Both the solid portion and cystic components were removed. The IAC was drilled open and the residual tumor within was also removed. Postoperative histopathological examination of the mass unexpectedly revealed the diagnosis of a meningioma, World Health Organization (WHO) grade I (Figure 2). No new neurological deficits were observed after the operation, and no signs of recurrence were found during the subsequent 24-month follow-up period.

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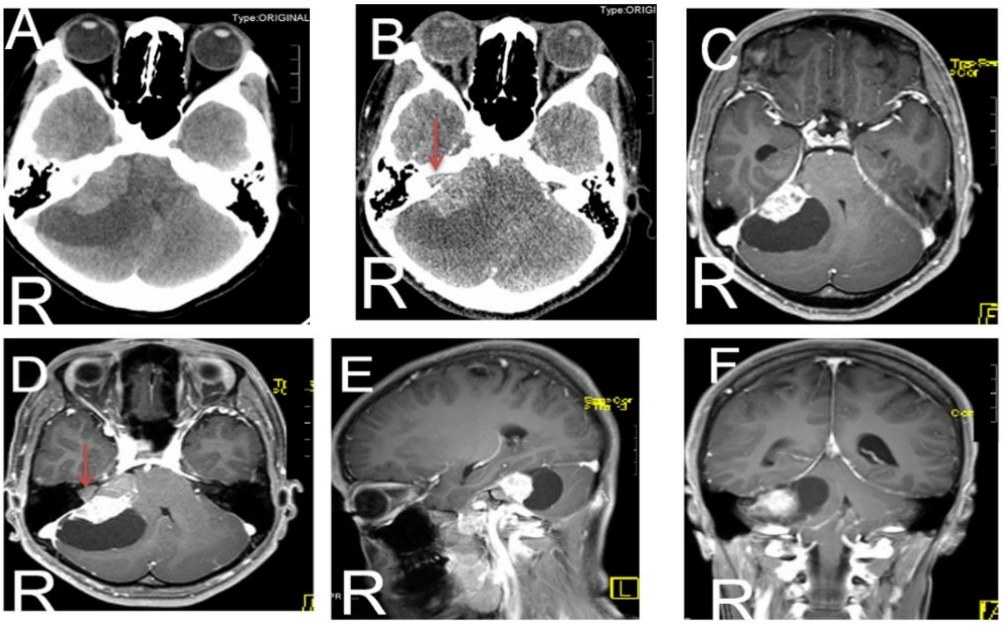

Figure 1: Cystic meningioma in CPA. **A)** Unenhanced head CT, **B)** enhanced head CT in axial reveal a cystic lesion with a broad base in the right cerebellopontine angle. **C)** & **D)** T1 weighted MR images with gadolinium in axial, **E)** sagittal and **F)** coronal plane show the cystic lesion is unevenly enhanced. **B)** & **D)** arrow, the solid part of the tumor protrudes into and enlarges the inner auditory canal.

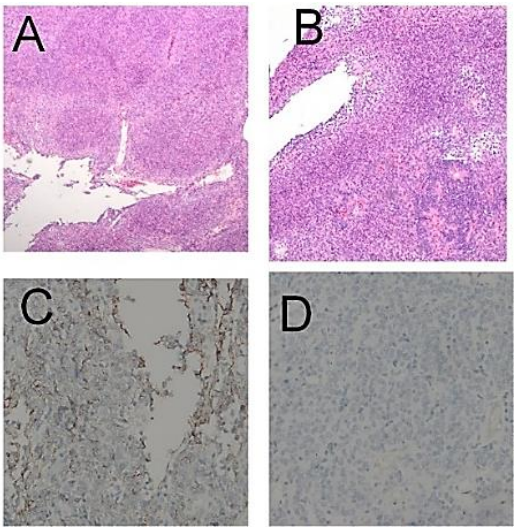

Figure 2: Pathological findings. Photomicrograph shows a cystic tumour consisting of meningothelial component (**A)** Hand E, ×40; **B)** H and E, ×100). Tumor cells show immunopositivity for EMA (**C)** EMA ×200) and immunonegativity for S-100 (**D)** S-100 ×200).

Discussion

Although meningioma is a fairly common intracranial tumor, cystic meningioma only accounts for 2% to 7% of all meningiomas [3, 4]. Most cystic meningiomas occur in the supratentorial region, especially in the frontal and parietal lobes [1]. At present, there are only 10 cases of CPA cystic meningioma in the English literature. The etiology, clinical features, surgical strategies and prognosis of the disease are not very clear. In this study, we reported a case of CPA cystic meningioma mimicking the vestibular schwannoma along with a literature review.

When it comes to CPA cystic lesions with enhanced nodules, the common preoperative diagnoses include vestibular schwannoma, metastatic tumor, and glioma, hemangioblastoma [1, 5-7]. Our findings suggest that cystic meningioma should be taken into consideration. Hemangioblastoma usually shows marked serpentine flow voids on MRI and vascularized nodules on CTA [8]. Gliomas often present heterogeneous enhancement patterns [3]. Metastatic tumors usually present with significant peripheral edema [9]. In this case, it is difficult to differentiate from vestibular schwannoma preoperatively because of the invasion of the lesion into the IAC and the enlargement of the IAC. However, the broad base of the tumor may be a giveaway sign. If the thickening of the adjacent dura, especially the typical dural tail sign, can be observed, it may be much easier to reach the diagnosis of cystic meningioma.

In 1979, Nauta *et al.* classified cystic meningiomas into four types according to the relationships among the cysts, the tumor, and the surrounding brain – Type I: intratumoral cyst centrally located in the tumor, Type II: intratumoral cyst lying at the edge of the tumor, Type III: peritumoral cyst lying in the adjacent brain, and Type IV: peritumoral cyst located between the brain and the tumor [10]. According to this classification, our case should be classified into type IV, which can be very misleading in terms of preoperative diagnosis. Different types of tumors are considered to be related to different pathogenesis of cyst formation. Peritumoral cysts are usually attributed to peritumoral edema, intratumoral hemorrhage, or cerebrospinal fluid retention. However, intratumoral cysts may be caused by ischemic necrosis, intratumoral hemorrhage, and increased secretion of tumor cells [3, 11]. Hui also attributes the formation of peripheral cysts to ultrafiltration of tumor vessels and increased tumor vascular permeability, especially considering that the nodular portion of cystic meningioma is often characterized by abundant blood supply [3]. Due to the scarcity of literature coverage, the exact pathogenesis of this phenomenon is still unknown. Further investigation is warranted.

A previous review of cystic meningiomas of CPA shows the incidence of clear cell type (WHO grade II) was 75% (9/12), the most common pathological type, followed by meningothelial subtype at 16.7% (2/12) [3]. So, complete resection, including the enhanced cyst wall, is the current standard treatment. However, there is no agreement as to whether the unreinforced cyst wall should be removed. Some believe that the unenhanced wall has no or few tumor cells, and therefore, it is not indispensable to remove it at the risk of increased bleeding [12, 13]. Others argue that the unreinforced cyst wall should be removed because it may have a small number of tumor cells [1]. The recurrence rate has been reported to be 9% in cases of cyst wall retention [14]. Hui suggests that intraoperative biopsy of the wall, postoperative adjuvant radiotherapy and close follow-up might be a better alternative.

Conclusion

Cystic meningiomas should be included in the differential diagnosis of CPA cystic lesions. Total resection of the tumors, including enhanced cyst wall, is the current standard treatment. Generally, their pathological subtypes are clear cell types, with a higher rate of postoperative recurrence. In light of this, the importance of close postoperative follow-up should be emphasized.

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Conflicts of Interest

None.

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