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Case Report

Glomus tumour masquerading as an aural polyp in chronic middle ear disease: A case report

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الملخص

ورم المستقتمات من الأورام غير الشائعة في العظمة الصدغية. ومن أعراضها المرضية الطنين النابض، والنزيف المتكرر من الأذن، وكذلك الصمم السمعي أو شلل في العصب السابع ونادرا ما يكون مصاحب بالتهاب قيحي مزمن في الأذن الوسطى. وفي هذه الحالة قد يعطي قراءة تشخيصية خاطئة لموجودات باثولوجيا الأنسجة.

نقدم حالة نادرة لسيدة عمرها ٤٥ عاما مصابة بورم كتبي في الأذن اليمنى، يصاحبه التهاب قيحي مزمن في الأذن الوسطى، مع بروز لحمية كبيرة الحجم من الأذن الخارجية. وبالرغم من إجراء الفحوصات اللازمة قبل العملية بما فيها الأشعة المقطعية إلا أنه لم يتم تشخيص الورم. وأثناء إجراء العملية الجراحية واجهتنا كمية كبيرة وغير معهودة من النزيف مما تسبب في إيقاف العملية بعد أخذ عينة من الورم. وقد كان التقرير المبدئي يوحي بوجود ورم كوليستيرولي، إلا أنه بعد مراجعة الشرائح مع وضع صبغات مناعية خاصة (S-١٠٠) تبين أن الحالة هي ورم المستقتمات. اللحميات المصاحبة لالتهاب الأذن الوسطى قد تخفي حقيقة هذا الورم الوعائي وبالتالي تتسبب في صعوبة التشخيص المجهري. نقترح أن يُضم الورم الكتبي إلى قائمة التشخيص التفريقي للحالات المشابهة. كما نقترح أنه عند وجود مؤشر عالي للاشتباه به يجب إجراء أقصى الفحوصات الشعاعية اللازمة المتوفرة قبل العملية وعمل الصبغات المناعية المناسبة.

الكلمات المفتاحية: الورم الكتبي؛ المستقتمات الرأسية الطبلية؛ تصوير؛ باثولوجيا الأنسجة

Abstract

Paraganglioma is an uncommon benign tumour of the temporal bones. It usually causes pulsatile tinnitus, recurrent ear bleeds, deafness or facial palsy and is rarely associated with chronic suppurative otitis media (CSOM). The latter may lead to false histopathological findings. We present an unusual case of a 45-year-old female with a right ear glomus tumour that was associated with CSOM and a large polyp protruding from the auditory canal. Despite preoperative investigations including computed tomography, diagnosis of the tumour could not be established. After taking a biopsy, a curative operation had to be abandoned because of a torrential intra-operative haemorrhage. The initial biopsy report suggested cholesteatoma; however, further histopathological studies including S-100 protein immune-staining revealed it to be paraganglioma. Large aural polyps and granulation tissues in CSOM can mask the characteristic histopathological features of these vascular tumours. We recommend including glomus tumour in the differential diagnosis of similar cases and performing optimum preoperative radiological investigations and immunological staining to confirm the diagnosis.

Keywords: Glomus tumours; Histopathology; Imaging; Jugulo-tympanic paraganglioma

Introduction

Jugulo-tympanic paragangliomas arise from small bodies of neuroendocrine tissue and are the most common slow-growing neoplasm arising from the middle ear.[1] The presenting features, usually in middle-aged women, include pulsatile tinnitus, recurrent ear bleeding, deafness, otalgia,

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dizziness and facial and lower cranial nerve palsies.[2] Though most temporal bone paragangliomas are diagnosed clinically and radiologically,[3] they are confirmed histopathologically by the presence of monomorphic cell nests, vascularized stroma and S-100 protein immuno-stains for chromogranin, synaptophysin and sustentacular cells.[2]

Indeed, difficulties may arise when these tumours masquerade as granulation tissue arising from the middle ear in patients with CSOM, which can only be diagnosed intraoperatively.[4]

Case presentation

A 45-year-old female presented to the otorhinolaryngology (ORL) outpatient department with a complaint of a recurrent right ear foul smelling discharge associated with deafness for more than two years.

Clinically, she had a large aural polyp protruding from the external ear canal with foul smelling mucopus discharge. Computerized tomography (CT) of the temporal bone reported a soft tissue filling the middle and external ear (Figure 1). A provisional diagnosis of chronic suppurative otitis media with possible cholesteatoma was made.

Intraoperatively and after cortical mastoidectomy, copious bleeding was encountered from the middle ear; taking the posterior canal wall down for better exposure did not help. Eventually, bone wax was used to control the bleeding, and the procedure was abandoned after taking a biopsy.

Postoperative investigations, including MRI (magnetic resonance imaging), indicated a vascular mass (Figure 2). Histopathology reported a cholesteatoma (Figure 3). Finally, S-100 protein and chromogranin stains confirmed a glomus tumour after combined surgical and radiological findings were discussed with the pathologist (Figure 4).

Discussion

The presenting features of this benign tumour are unique; thus, the initial clinical diagnosis can be fairly accurate. Moreover, it can be confirmed preoperatively by certain imaging techniques including MDCT (multidetector computed tomography), MRI, CT angiogram and angiography when embolization is planned.[5,6]

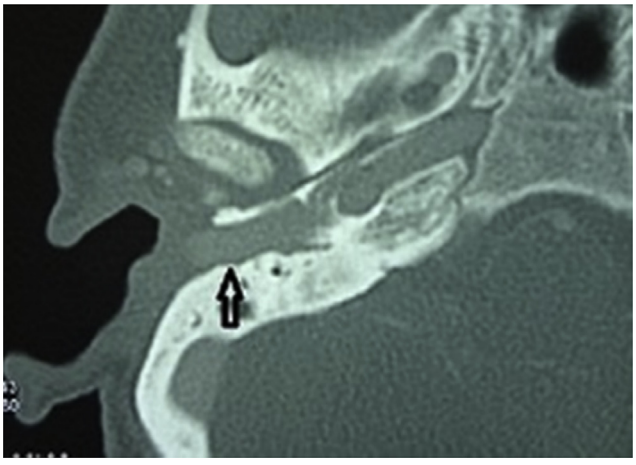

Figure 1: Preoperative CT of the right temporal bone demonstrating an aural polyp filling the external auditory canal (black arrow).

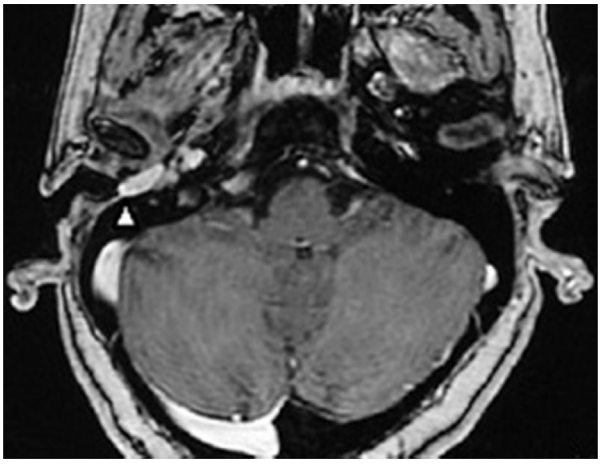

Figure 2: Postoperative MRI of the right temporal bone with contrast demonstrating the aural polyp remnant (arrow head).

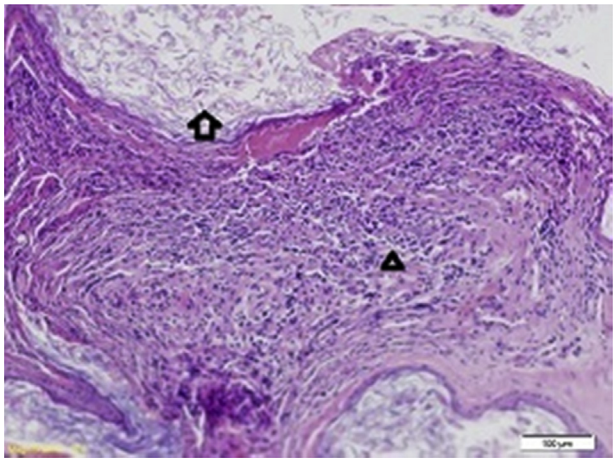

Figure 3: (H&E × 200) Sections show keratinizing epithelium with keratin flakes representing cholesteatoma (arrow) and the underlying paraganglioma (arrow head).

However, granulation tissue and/or an aural polyp in CSOM appear enhanced in a CT scan with gadolinium diethylene-triamine-pentaacetic acid (DTPA), causing cognitive difficulties or misdiagnosis when associated with glomus tumours,[7,8] and the diagnosis can only be made postoperatively.[9]

Confusion may also increase when the tissue that is extracted does not represent the actual pathology because of a long-standing exposure that could distort the clinical, gross and microscopic findings; this may explain the pathologist's initial report, as seen here.

Interestingly, most of the treatment options reported for this entity, including stereotactic radiosurgery, radiotherapy, chemotherapy and intramural sclerosing agent, metabolic therapy with I^{131} [10] and surgical excision, do not require histopathological specimens.

Nevertheless, it is fair to admit, with justification, that the presentation and the limited preoperative investigations done for our patient unavoidably missed the diagnosis for the reasons given above.

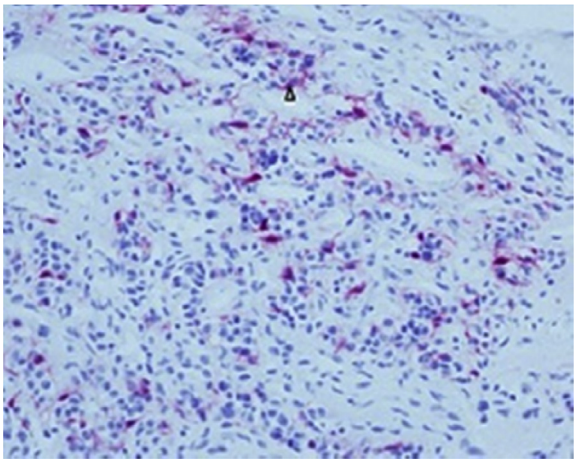

Figure 4: Immunohistochemistry stain of sustentacular cells of paraganglioma highlighted by S-100 stain (arrow head).

For similar cases with a high index suspicion of glomus tumour, we recommend the optimum radiological investigation of the available choices.

There may be an argument regarding the cost effect of the investigations suggested; nonetheless, in these low incidence cases, considering the risk of intraoperative untoward findings, we believe it is justified.

Conclusion

Paraganglioma can be associated with chronic ear disease that masquerades its presentation. We recommend including glomus tumour in the differential diagnosis in similar cases with a high index of suspicion and performing optimum radiological investigations and immunological staining in order to confirm the diagnosis.

Sponsorship

None.

Conflict of interest

None.

Authors' contribution

SAA is the sole contributor of the article. He conceived and designed the study, conducted research and organized data. Also drafted the article, finalized it and responsible for the content and similarity index of the manuscript.

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