Introduction

Carcinoid tumor is a neuroendocrine tumor traditionally from the gastrointestinal tract and lung [1] and is rarely found in the middle ear (ME). Occasionally, such cases are also reported as primary tumor in the testes [2]. ME carcinoid tumor was first reported in 1980 by Murphy et al [1]. Patients with carcinoid tumor of the ME usually present with nonspecific symptoms including conductive hearing loss, aural fullness, and less commonly tinnitus, otalgia, and headache with acute facial palsy a rare finding. ME exploration is necessary for the common differential diagnoses of a mass in the ME with acute facial palsy, which should include cholesteatoma, glomus tumor, carcinoma, facial nerve neroma, and other tumors. ME carcinoid tumor is easily confused with ME adenoma by histopathology alone, and the definitive diagnosis is made by immunohistochemistry and ultrastructural examination of biopsy materials, and its treatment of choice is radical resection of the tumor with a wide margin by canal wall-down approach[3,4].

Case Report

A 30 year old lady presented with right ear bleed and otalgia of one month duration. She also described progressive right tinnitus and hearing loss a year preceding this episode accompanied by facial palsy of 4 months duration. No related history of fever and auricular vesicles was present nor were there symptoms of carcinoid syndrome, such as flushing, wheezing, cramping and diarrhea. A reddish polypoidal mass was observed during otoscopy and right facial palsy was staged as grade V on the House-Brackmann scale. Initial audiogram showed normal hearing in the left ear and a 90-dB hearing loss in the right ear at 500dB, 1000dB, 2000dB and 4000dB. As the patient was at her 37th week of pregnancy at the time of the presentation, imaging was delayed till after delivery. After delivery, the high resolution computed tomography (HRCT) of the temporal bone done showed soft tissue opacification of the external acoustic meatus (EAM), middle ear and mastoid air cells with rarefaction and destruction of the right petrous part of temporal bone apex. However, the patient defaulted her clinic appointment for two years and experienced worsening of symptoms. A repeated HRCT showed further extension of the temporal bone destruction predominantly involving the petrous part with erosion of the tegmen tympani. A magnetic resonance imaging (MRI) was performed with similar findings of the HRCT whilst showing no dural involvement (Figure 1).

Under the suspicion of ME tumor, a lateral resection of the right temporal bone was performed. Tumor mass biopsy sent for histopathological examination after the operation was...
reported as ME carcinoid tumour. The postoperative tumor surveys for distant metastases were unremarkable, and patient was referred to the oncology side for radiotherapy.

The postoperative course was uneventful. Right otalgia improved immediately after the operation. However, right facial palsy and degree of hearing loss persisted. Follow-up clinical examination and HRCT of temporal bone revealed neither signs of local recurrence nor regional metastasis.

Discussion

The middle ear (ME) rarely presents with primary neoplasm and carcinoid tumors are amongst the most uncommon occurrence of these infrequently seen tumors [5]. This case shows the challenge faced in diagnosing such cases. In fact, ME carcinoids may be under reported as it is challenging to distinguish it from other ME adenomatous tumors by histopathology [6]. Similar to most benign or nonaggressive middle ear tumors, our patient presented with nonspecific symptoms including hearing loss, aural fullness, tinnitus, otorrhea, and otalgia, although facial palsy and/or regional metastases was evident in some of the patients [5]. The case reported showed a slowly progressing tumour as reported in other reviews. ME carcinoid appears to follows a less aggressive course compared to carcinoid tumors elsewhere in the body [7], yet studies have shown cases where regional lymph node metastasis indicate that it is not exactly an innocuous neoplasm. Similar to the case reported, local invasion is habitually slow and nondestructive. Invasion into the external auditory canal, eustachian tube, inner ear, mastoid and consequent facial palsy have been reported as seen in the case we encountered [5]. Incomplete and conservative excisions have shown local recurrences, some of which can be accepted as residual tumor [8]. However, a proportion of cases have recurred even with radical operations [9].

Based on histopathology alone, it is difficult to distinguish between ME carcinoid tumor and other ME adenomatous tumors such as adenoma [6]. With routine light microscopy, both are composed of small uniform cells arranged in glandular, trabecular, and solid patterns. However, ME carcinoid tumor cells are immunohistochemically reactive to antibodies to chromogranin A, neuron-specific enolase, keratin, and synaptophysin, while ME adenoma are not. Furthermore, the electron microscopy would demonstrate numerous neurosecretory granules in the carcinoid tumor cells (Figure 2).

The possibility of regional metastasis is extremely low but radiotherapy was suggested in our patient due to the delayed treatment. This was subsequently supplemented with regular follow-ups with clinical examination and imaging studies. Local recurrences after complete excision is rare, and a more extensive surgery like extended subtotal petrosectomy may be required if it eventualize [9].

Conclusion

A degree of clinical suspicion is required when encountering a growing mass that originated from behind the tympanic membrane with nonspecific symptoms, to consider middle ear adenomas, and carcinoid tumors. Definitive diagnosis can be made using histopathology of the biopsy materials. Due to its rare occurrence, reporting the clinical presentation and features of each new case would help to delineate the characteristics and therefore, increasing the awareness towards the disease.

Figure 1: Immunohistochemically stained histological images if carcinoid tumour. Neoplastic cells are positive for chromogranin A. (magnification, x100).

Figure 2: Axial view of the MRI Brain showing destructive soft tissue lesion predominantly involving the petrous part of the right temporal bone with extension into the middle and inner ear and the internal auditory meatus.

Characteristics of carcinoid tumor of the middle ear

1. Rare
2. Delay in diagnosis is common
3. Slow growing tumor
4. Definitive diagnosis can be made using histopathology
5. High suspicion and awareness of the disease is required

References

1. Murphy GF, Pilch ZB, Dickersin GR, Goodman ML, Nadol JB (1980) Carcinoid tumor of the middle ear. Am J Clin Pathol 73: 816-823. Link: https://goo.gl/FLqL3c
2. Bolat MS, Akdeniz E, Saltik F, Sahinkaya N, Moral C (2015) Primary Carcinoid Tumor of the Testis: Case Report. Urol Case Rep3: 152-154. Link: https://goo.gl/6qHP9
3. Manne JJ, van Haelst UJ, Kubat K, Marres EH (1984) Primary carcinoid of the middle ear. A case report with reference to light and electron microscopy findings. Hno 32: 419-423. Link: https://goo.gl/KfSHgF

4. Krouse JH, Nadol JB, Goodman ML (1990) Carcinoid tumors of the middle ear. Ann Otol Rhino Laryngol 99: 547-552. Link: https://goo.gl/e5Dbkj

5. Ahan M, Yildirim N, Arslanoglu A, Karşıoğlu Y, Kazıkdas KÇ (2008) Carcinoid tumor of the middle ear: report of a case 29: 352-356. Link: https://goo.gl/QSJuQv

6. Shibosawa E, Tsutsumi K, Ihara Y, Kinoshita H, Koizuka I (2003) A case of carcinoid tumor of the middle ear. Auris Nasus Larynx 30: 99-102. Link: https://goo.gl/SeYHdW

7. Soga J (2003) Carcinoids and their variant endocrinomas. An analysis of 11842 reported cases. J Exp Clin Cancer Res 22: 517-530. Link: https://goo.gl/DZPxVi

8. Ramsey MJ, Nadol JB, Pilch BZ, McKenna MJ (1960) Carcinoid tumor of the middle ear: clinical features, recurrences, and metastases. Laryngoscope 115: 1660-1666. Link: https://goo.gl/RbJ96k

9. Knerer B, Matula C, Youssefzadeh S, Ulrich W, Swoboda H (1998) Treatment of a local recurrence of a carcinoid tumor of the middle ear by extended subtotal petrosectomy. Eur Arch Otorhinolaryngol 255: 57-61. Link: https://goo.gl/MsBbjQ