Clinical Case Report

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Facial nerve schwannoma mimicking chronic suppurative otitis media

A case report

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Abstract

Rationale: Facial nerve schwannoma (FNS) is a rare slow-growing nerve sheath tumor derived from Schwann cells. FNS with normal facial nerve function may sometimes be misdiagnosed as otitis media because of similar ontological symptoms such as purulence, tympanic membrane damage, and hearing loss.

Patient concerns: A 68-year-old woman was referred to our department because of otorrhea and hearing loss in the right ear for 20 years. Otoscopy revealed abundant purulent secretions deep in the right external auditory canal, and granulation proliferation in the posterior part of membranae tensa. Audiogram showed a right mixed hearing loss with an 85-dB pure-tone average and 35-dB air-bone gap.

Diagnosis: This patient was misdiagnosed as chronic suppurative otitis media before surgery. During surgery, a mass was found, and intraoperative frozen section histopathology confirmed an FNS.

Interventions: This patient was subjected to mastoidectomy for curing chronic suppurative otitis media initially. During surgery, a mass was found attached and widely extended into the tympanic and mastoid segments. We removed most part of the mass, however found the mass deriving from the vertical part of the facial nerve. Intraoperative frozen section histopathology confirmed an FNS. So we removed the incurs and malleus, and searched for the edge of the mass. The mass involved multisegments of facial nerve including the tympanic, vertical and pyramidal segments. The tumor was removed completely, and nerves were repaired using greater auricular nerves.

Outcomes: After surgery, the patient had facial nerve paralysis of House-Brackmann (HB) Grade VI. Facial function recovered to HB Grade III at 30 months after surgery. The patient was followed up for 5 years. She had a facial function of HB grade III at the most recent follow-up.

Lessons: FNS is rare and tend to be misdiagnosed. It is important to combine the imaging modalities of computed tomography and magnetic resonance imaging to evaluate FNS before surgery. The primary goal of managing FNS is to maintain normal facial function as long as possible; therefore, tailored strategy should be taken for managing FNS.

Abbreviations: ABG = air-bone gap, CT = computed tomography, EAC = external auditory canal, FNS = facial nerve schwannoma, HB = House-Brackmann, MRI = magnetic resonance imaging, PTA = pure-tone average.

Keywords: chronic suppurative otitis media, facial nerve schwannoma, facial paralysis, hearing loss, misdiagnosis

1. Introduction

Facial nerve schwannoma (FNS) is a rare slow-growing nerve sheath tumor derived from Schwann cells.[1] FNS constitutes only 0.15% to 0.8% of allintratemporal tumors.[2] FNS can arise at any segments of the facial nerve varying from the cerebellopontine angle segment to the temporal outer segment. Most FNS locate at the geniculate ganglion which derives from the sensory nerve fiber of the facial nerve. The most common presentation of FNS is facial paralysis, followed by sensorineural or conductive hearing loss, tinnitus, nostalgia, and so on.[3] FNS is easily to be misdiagnosed because of variable and nonspecific clinical presentations. Particularly for those cases with normal facial nerve function, FNS may sometimes be misdiagnosed as otitis media because of similar ontological symptoms such as purulence, tympanic membrane damage, and hearing loss. Current management of FNS is also challenging with debating therapy strategies.

Herein, we report the clinical presentations, diagnosis, treatment, and outcome of a patient with FNS misdiagnosed as chronic suppurative otitis media.

2. Case report

A 68-year-old woman was referred to our department because of otorrhea and hearing loss in the right ear for 20 years. Otoscopy examination revealed abundant purulent secretions deep in the

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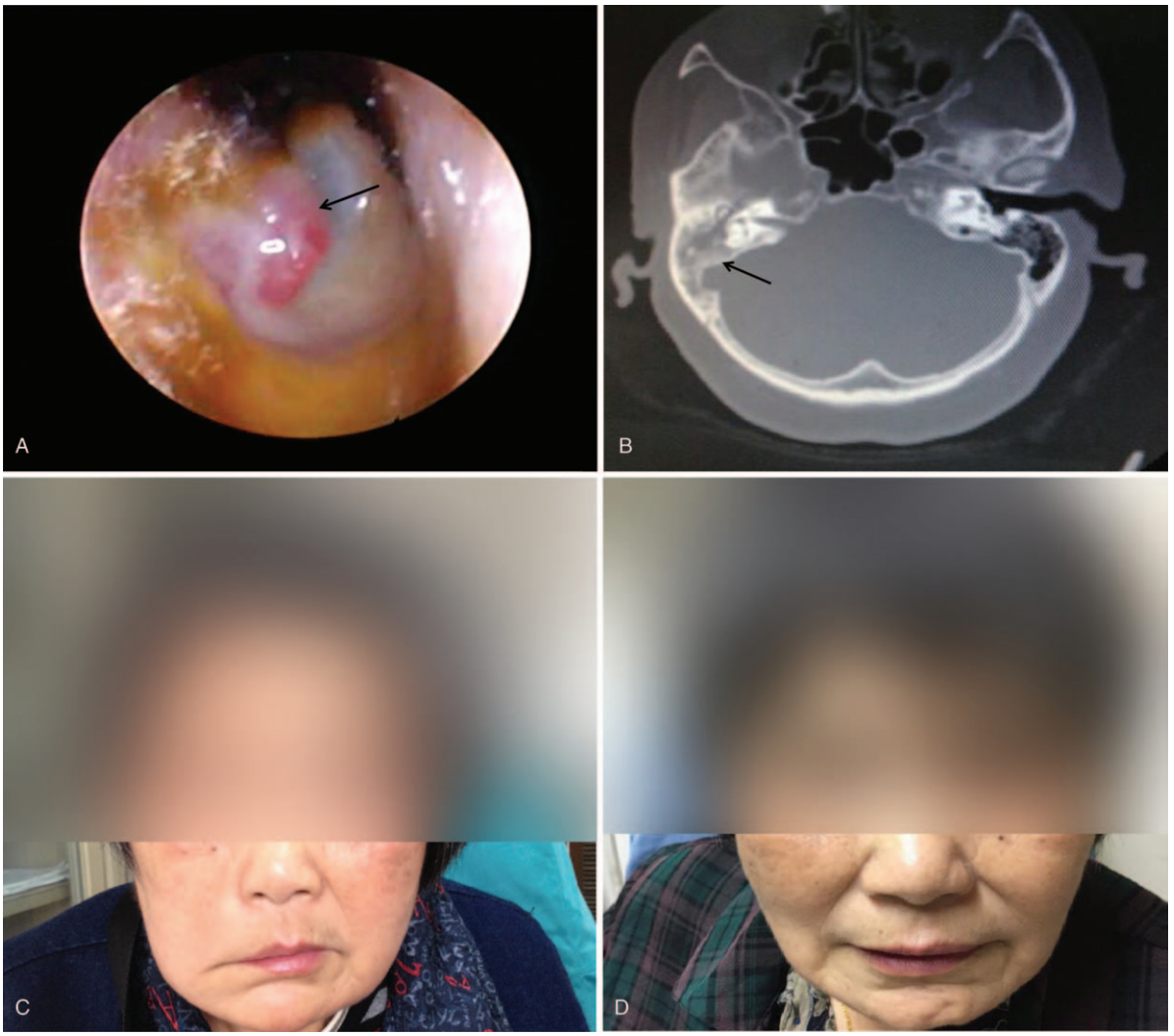

Figure 1. (A) Otoscopy examination showing abundant purulent secretions deep in the right external auditory canal, and granulation proliferation in the posterior part of membranae tensa. (B) Computed tomography scanning showing the tumor presenting in the tympanic and mastoid segments of right ear. (C) The patient had facial nerve paralysis of House-Brackmann Grade VI. (D) Facial nerve function improved to House-Brackmann Grade III at 30 months after surgery.

right external auditory canal (EAC), and granulation proliferation in the posterior part of membranae tensa (Fig. 1A). Audiogram showed a right mixed hearing loss with a 85-dB pure-tone average (PTA) and 35-dB air-bone gap (ABG). Preoperative computed tomography (CT) showed a soft tissue shadow filling the mastoid cavity and tympanic sinus. The patient had normal facial nerve function, so she was diagnosed with chronic suppurative otitis media based on the imaging evaluation and otoscopy examination before surgery. We proposed to perform mastoidectomy to cure chronic suppurative otitis media for the patient. During surgery, we found a mass attached and widely extended into the tympanic and mastoid segments (Fig. 1B). There was no clear demarcation between the edge of the mass and normal nontumorous tissue. We removed most part of the mass, however found the mass deriving from the vertical part of the facial nerve when cleaning up the remaining tissue. Intraoperative frozen section histopathology also confirmed an FNS.

Then we removed the incurs and malleus, and searched for the edge of the mass. The mass involved multisegments of facial nerve including the tympanic, vertical, and pyramidal segments. The tumor was then removed completely, and nerves were repaired using greater auricular nerves. After surgery, we re-reviewed CT imaging of the patient and found a suspected enlargement of bony canal in the conical segment of the vertical facial nerve. After surgery, the patient had facial nerve paralysis of House-Brackmann (HB) Grade VI (Fig. 1C). Facial function recovered to HB Grade III at 30 months after surgery (Fig. 1D). The patient was followed up for five years. Facial function was found to HB grade III at the most recent follow-up. Hearing loss was not improved, and audiogram showed a severe sensorineural hearing loss with 90 dB-PTA.

The study was approved by the Ethics Committee of Affiliated Hangzhou First People's Hospital, Zhejiang University School of Medicine. A written informed consent was obtained from the patient for reporting the case details as well as the pictures.

3. Discussion

FNS is a rare tumor difficult to be diagnosed and managed. FNS is often multi-segments involved and locates at the labyrinthine and the geniculate ganglion.[4] Facial weakness is the most frequent

symptom, followed by hearing loss. However, facial nerve function is normal in some patients because the tumor grows very slowly and the facial nerve compression can be tolerated. Sunderland et al[5] reported that a normal facial nerve function occur in 27.3% of all patients with FNS. For the very rare cases with mass eroding through the posterior bony wall of the externalauditory canal, 27% to 50% of patients have normal facial function[6]; these patients are often misdiagnosed as middle ear diseases. The reasons of misdiagnosis for our case were as following: the patient only presented with suppurative otitis media and hearing loss however without facial nerve paralysis or weakness even during such a long period of 20 years; preoperative CT scanning only showed a soft tissue shadow filling the mastoid cavity and tympanic sinus, and suggesting suppurative otitis media. The lesson learned from this case was that pre-operative MRI should be performed. Preoperative MRI may be helpful in the diagnosis of FNS in our case. FNS may show moderately enhanced MRI imaging; however, otitis media shows normal MRI signals. Quesnel et al[7] suggested that CT and MRI are critical for correct diagnosis of FNS.

Because of the rarity of these tumors and the benign and slow-growing nature, still no universally accepted management is avaliable.[8] Therapeutic options of FNS depends on facial function, tumor size, and evolution and disabling symptoms such as hearing, dizziness, and so on.[3] Therapeutic strategies for FNS include surgical intervention, observation, and radiotherapy of schwannomas to preserve facial function for a longer period of time.[8] Surgical resection with facial nerve repair is usually the standard management for patients with facial nerve function of HB grade III or worse.[9] For patients with normal facial nerve function, a "wait and see" strategy and nerve-preserving technique is recommended.[7,8,10,11] Park et al[12] reported that the proximal portion of the geniculate ganglion and small tumors (<2 cm) involving 1 or 2 facial nerve segments are the best candidates for nerve-sparing stripping surgery. For our case, misdiagnosis leads to wrong management strategy. When performing mastoidectomy, we resected the mass which leading to the cut-off of partial normal facial nerve. Although the facial nerve was reconstructed, facial nerve function deteriorated from normal before surgery to be HB grade VI after surgery. If diagnosed correctly before surgery, "wait and see strategy" will be adopted for this patient.

In conclusion, FNS is rare and difficult to be diagnosed and managed. It is important to combine the imaging modalities of CT and MRI to evaluate FNS before surgery. The primary goal of managing FNS is to maintain normal facial function as long as possible; therefore, tailored strategy should be taken for managing FNS.

Author contributions

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