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Unilateral vestibular schwannoma associated with a Jacobson's schwannoma

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Abstract. – Coexistence of unilateral vestibular schwannoma and Jacobson's schwannoma growing in the same intracranial site is rarely observed.

We present the case of 36-year-old woman with primary diagnosis of vestibular schwannoma and subsequent appearance of schwannoma to the Jacobson's nerve. Initial wait and see strategy was performed offered us the opportunity to describe Jacobson's lesion features at computed tomography over a period of 4 years. Subtotal petrosectomy with infralabyrinthine approach was subsequently executed to remove the growing mass of the temporal bone.

The Jacobson's schwannoma increased its size from 0.4 cm for years whereas vestibular schwannoma size was unchanged after 7 years observation.

The concomitant removal of both schwannomas is still associated with the size of the CPA lesion and to patient's symptoms.

Key Words:
Multiple schwannoma, Jacobson's schwannoma, Vestibular schwannoma.

Introduction

The occurrence of multiple schwannomas growing in the same intracranial site is rarely observed, except in patients affected by neurofibromatosis types 1 (NF-1) and 2 (NF-2)[1,2].

We present the case of a patient, not affected by NF, suffering from two asynchronous schwannomas, emerged to the vestibular nerve and to Jacobson's tympanic one. There are few reports in the literature about Jacobson's nerve (JN) schwannoma in the middle ear and temporal bone[3-7]. To our knowledge, no author has previously reported the coexistence of these two tumors in the same patient. Moreover, this report is the first to describe the Jacobson's lesion features at computed tomography (CT) over a period of 4 years.

Case Report

In July 2005, a 36-year-old woman came to our Department complaining severe bilateral hearing loss. Otoscopy was normal and pure tone audiometry showed bilateral sensorineural hearing loss. Gadolinium-enhanced MRI was planned (Figure 1 A,B,C). MRI revealed soft strongly enhancing tissue in the left internal auditory canal (IAC). We suspected a left vestibular schwannoma (VS). Wait and see strategy were initially followed.

Due to her bilateral progressive profound hearing loss the patient underwent right cochlear implant in 2007.

In January 2008, the patient complained a left slight ear fullness with an otoscopic appearance of a mass behind the tympanic membrane. CT were planned because cochlear implant contraindicated MRI testing. This latter showed the middle ear partially obliterated by small soft tissue mass involving the promontory and the round window niche. No erosion of the cochlear promontory was seen. Maximum diameter of the mass in the middle ear was 0.4 cm (Figure 2 A,B). The size of the IAC mass was stable. Surgery was recommended, but the patient refused the surgical intervention.

Control CT were carried out in 2009 and 2010. In February 2009 the soft tissue mass was further involving the hypo and mesotimpanum, still confined to the tympanic cavity (Maximum diameter 1 cm) (Figure 3 A,B). In April 2010 the mass was expanded in the epytimpanum and anterior mesotimpanum and erosive changes in the posterior aspect of the mastoid was evident. Maximum

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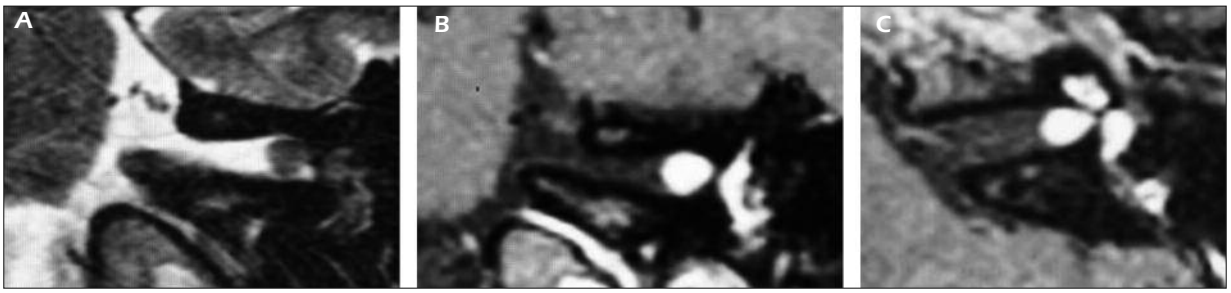

Figure 1. ***A-C,*** MRI coronal plane T1-W images: soft strongly enhancing tissue post contrast in the left internal auditory canal, indicative for VS.

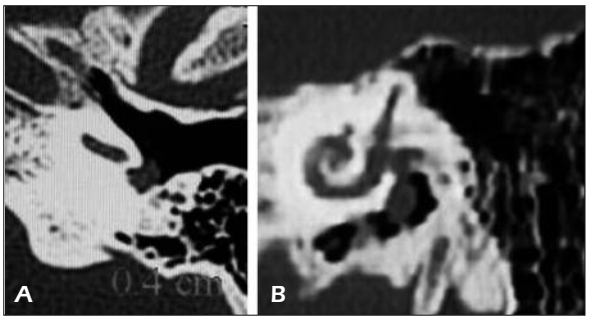

Figure 2. ***A, B,*** CT, axial plane Small soft tissue mass involving the promontory and the round window niche. Maximum diameter of the mass in the middle ear is 0.4.

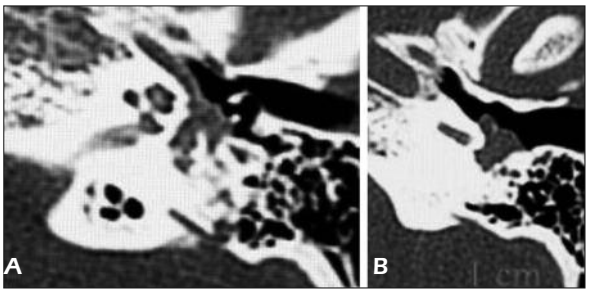

Figure 3. ***A, B,*** CT, axial plane. Soft tissue mass involving the hypo and mesotimpanum. Maximum diameter of the mass in the middle ear is 1.0 cm.

diameter in the middle ear increased to 1.6 cm (Figure 4 A,B,C,D).

In January 2012, the patient returned to our Institute with a left partial facial paralysis involving the inferior lips (grade III House-Brackman classification). A new TC scan showed maximum diameter of the mass had become about 2.5 cm occupying almost completely the mastoid and reaching the dura of the posterior cranial fossa (Figure 5 A,B,C). Again no growth for IAC mass could be detected.

At this time subtotal petrosectomy with infralabyrinthine approach was performed to remove the growing mass of the temporal bone. It arised from the Jacobson's nerve at the inferior portion of the promontory. We were able to remove the entire lesion preserving the anatomical integrity of the dehiscent facial nerve. Postopeartive histology confirmed the diagnosis of Jacobson's nerve schwannoma (JS). No middle ear recurrence and no CPA (cerebello pontine angle) lesion growth were observed after 1 year follow-up. After 4 months facial nerve function recovered reaching the grade II of the House-Brackmann classification.

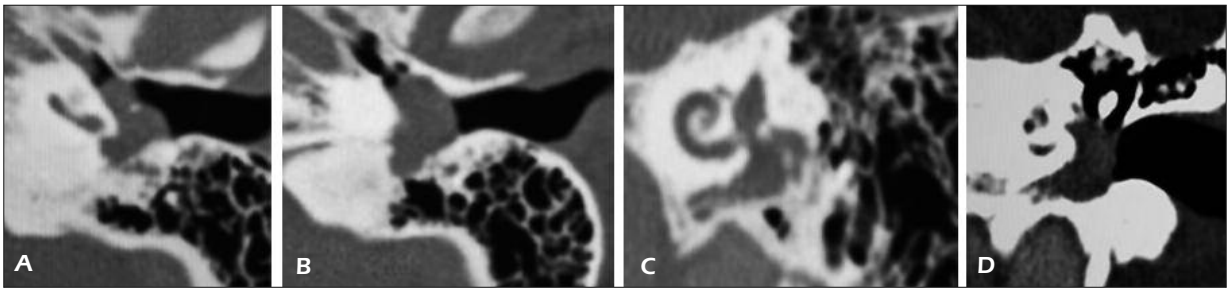

Figure 4. ***A-D,*** CT, axial plane. Soft tissue mass expanded in the epytimpanum and anterior mesotimpanum. Erosive changes in the posterior aspect of the mastoid are evident. Maximum diameter of the mass in the middle ear is 1.7 cm.

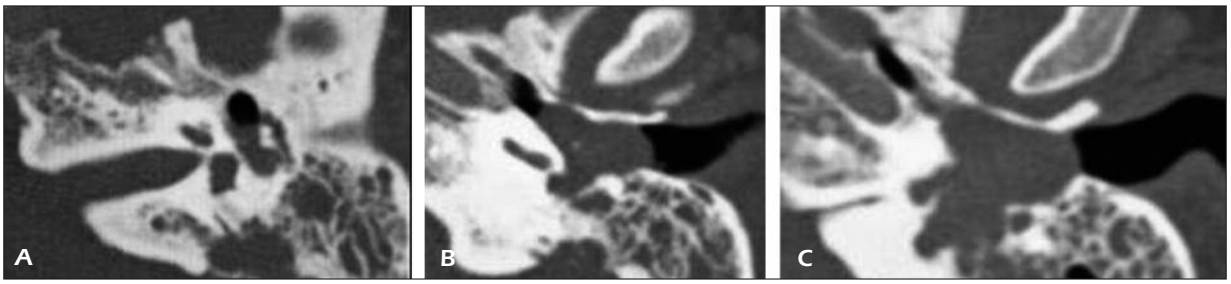

Figure 5. ***A-C,*** CT, axial plane. Maximum diameter of the mass about 2.5 cm with massive involvement of the mastoid and reaching the dura of the posterior cranial fossa.

Discussion

Usually multiple schwannomas develop in patients with genetically transmitted diseases such us NF-1 and NF-2 or after a repeated radiation exposure[1,2].

Just few cases of isolated Jacobson's neurinoma have been described[3-7]. Although there are published in literature VS associated ipsilaterally or contralaterally with other cranial nerves[2], no papers reported the coexistence of JS with VS.

In our patient, MRI confirmed the VS but subsequently we could not employ it again due to the cochlear implant insertion[8].

The majority of the middle ear schwannomas originate from the facial nerve. Other available neurogenic source of Schwann cells susceptible to tumoral development are chorda tympani, Arnold's nerve and Jacobson's nerve[3-5].

Aydin et al[3] gave as essential criteria for imaging diagnosis of JN the presence of erosion of the cochlear promontory and enlargement of inferior canaliculus. In contrast, Vasama et[5] al did not observe in their case such imaging clues and they based the radiological diagnosis on the absence of involvement of surrounding structures (jugular foramen, carotid canal and mastoid cells). However this tumor may grow invading temporal bone and skull base. CT scans are further characterized by the presence of a well circumscribed mass possibly extended in the mesotympanum, hypotympanum and/or epitympanum with concomitant erosion of one or more ossicles of the ossicular chain.

Further, it is very difficult perform a differential diagnosis between JS and facial nerve schwannoma on MRI[4,6]. They both appear typically isointense on T1 and T2 weighted images with a marked and homogeneous contrast enhancement.

Our CT data demonstrated initially a 0.4 cm soft tissue mass involving the promontory and the round window niche, but they did not show neither the erosion of the cochlear promontory nor enlargement of the inferior canaliculus in the early phases of growth lesion.

The request of the patient to apply a wait and see strategy offered us the opportunity to follow-up the evolution of the schwannoma. To our knowledge, this is the first report that has studied this aspect of Jacobson's schwannoma. The tumor increased its size from 0.4 cm calculated in 2008 up to 2.5 cm of the 2012. This means a growth of 0.4 cm per year. This growth rate is comparable with that documented in the evaluation of this factor in vestibular schwannoma[9]. The 2012 CT, confirmed at surgery, testifies that the mass, initially confined to the tympanic cavity, had involved the jugular foramen, the tympanic segment of the facial nerve, the mastoid and dura of the posterior cranial fossa. Pneumatic mastoid of the patient not representing a strong barrier to obstacle the expansion of the tumor probably favoured this aggressive behaviour. It is very interesting to note the remarkable JS extension into the neighboring areas otherwise to the growth lack of VS.

Surgery have not to be postponed in the event that the tumor is not confined to the middle ear or when facial nerve is involved[6-7]. Clinically, the outcome of surgical resection of a JN schwannoma is likely to be more favorable of facial nerve schwannomma because the facial nerve palsy can be spared[5-7]. The concomitant removal of both schwannomas is still associated with the size of the CPA lesion and to patient's symptoms.

Conflict of Interest

The Authors declare that they have no conflict of interests.

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