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Original Research

Jugulo-Tympanic Paraganglioma: Case Report

Ouziane Megherbi[1] | Yassine Merad[1] | Adel Tahraoui, Sarah Lazib[1] | Nabila Dechache[1]

[1]Universitary Hospital of Sidi Bel Abbes. Algeria

Abstract:

Paragangliomas, which frequently affect the head and neck, are usually benign, vascular tumors.

We report on 82-year-old female, with a medical history of diabetes mellitus type 2 complaining of pulsating tinnitus of the right ear and headache during one year. Otoscopy revealed retrotympanic pulsatile purple tumor of the right ear.

CT scan revealed an osteolytic tumor at the base of the skull that had reached the jugular foramen and was eroding the petrous bone. MRI displayed a tumor measuring 29 x 27 mm, on a height of 57 mm, with a "salt and pepper" pattern of hypo intensity. Given the large intracranial extension, the age and the clinical history of the patient, the patient underwent radiotherapy in replacement of surgery.

Introduction:

Jugulo-tympanic paragangliomas are the most common paragangliomas of the head and the neck area; they are the most frequent tumors of the petrous bone. Despite of their neuroendocrine origin, very few tympanic and jugular paragangliomas (1 to 3%) are secreting[1]

Most often benign tumors, there are sporadic and familial forms linked to mutations in the enzyme succinate dehydrogenase. Clinical presentation can vary across a wide spectrum of signs and symptoms. Therefore, the diagnosis rely on clinical signs and specifically on imagery results. Treatment is not consensual.

Case report:

We report on 82-year-old female, with a medical history of diabetes mellitus type 2, associated to high blood pressure; she was complaining of pulsating tinnitus of the right ear and headache during one year.

Otoscopy revealed retrotympanic pulsatile purple tumor of the right ear (Fig. 1). The neurological examination find lower cranial neuropathies (IX–XII) associated to a diplopia (VI). However, there were no intracranial hypertension on ophthalmological examination.

Pure tone audiometry showed a mixed hearing loss (Fig. 2)

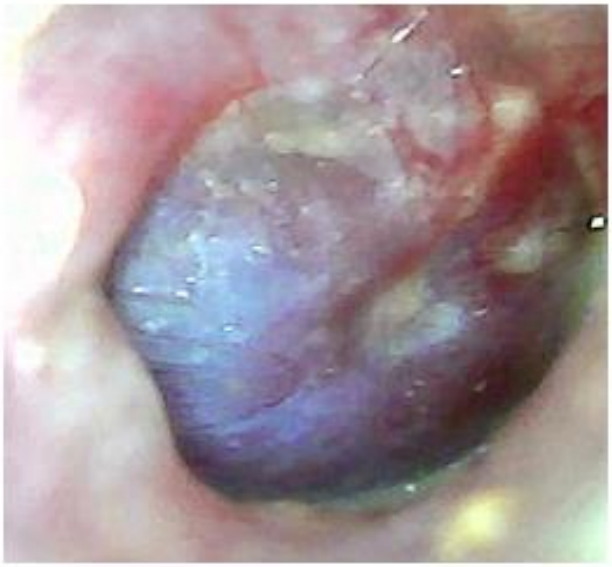

Fig. 1. Otoscopy image

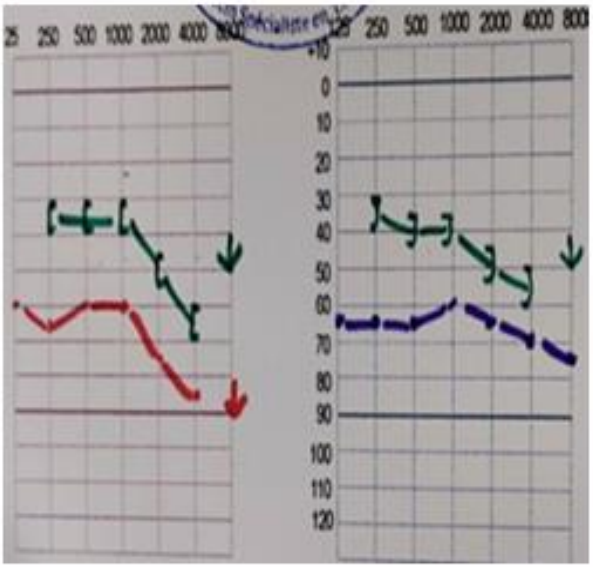

Fig. 2. Audiometry result

CT scan showed an osteolytic tumor of the skull base, eroding the petrous bone and reaching the jugular foramen (Fig. 3)

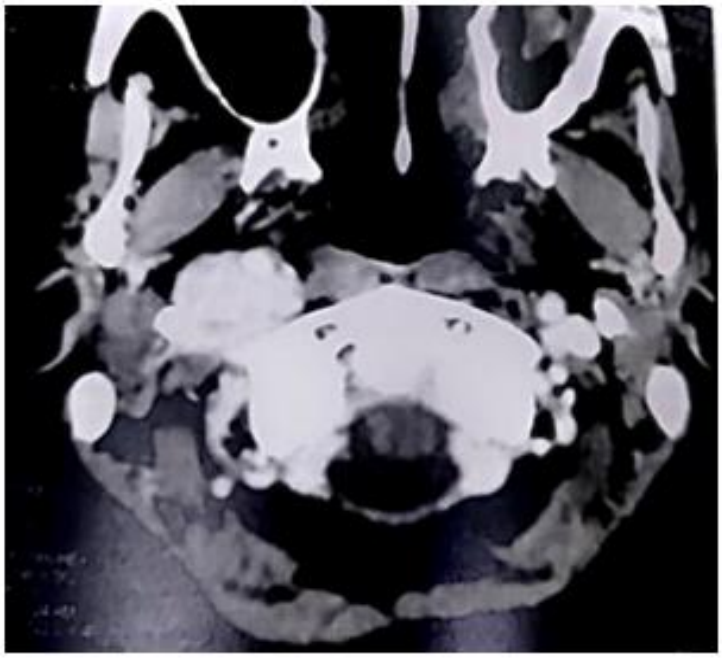

Fig. 3. CT scan image

A high resolution MRI demonstrated a mass filling a large part of the right temporal bone measuring 29 x 27mm, on a height of 57mm, with a "salt and pepper" pattern of hypo-intensity and hyper-intensity on T1-weighted images, relatively higher in intensity on T2-weighted images, and strongly enhanced after gadolinium injection. The MRI displayed an intra-cranial extension (cerebellous fossa) involving vascular and nervous structures of the cerebellopontine angle (Fig. 4).

Biological tests has eliminated a secreting paraganglioma variety Given the large intracranial extension, the age and the clinical history of the patient, the patient underwent radiotherapy in replacement of surgery.

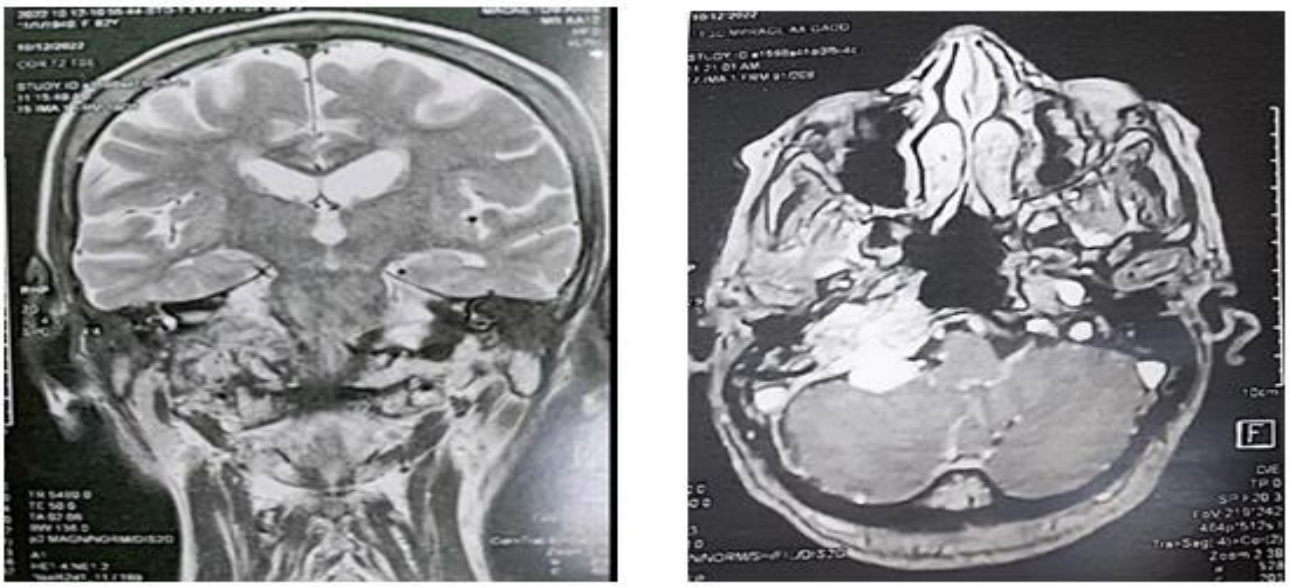

Fig. 4. MRI images

Discussion:

Tympanic and jugular paragangliomas are benign tumors developed from the neuroectodermal cells of the tympanic cavity and the dome of the jugular vein

In Erickson´s series of 204 head and neck paragangliomas, 57% were localized in the carotid body, 30% were tympano-jugular and 13% vagal [2].The literature mentions that females are more prone to developing this tumor[1].

The most frequent symptom reported in the literature is pulsating tinnitus. Progressive unilateral hearing loss could be explained by an ossicular lysis or a cochlear invasion. Less frequent manifestations are otalgia, vertigo, and neurological deficits (hoarseness, dysphagia and facial nerve palsy)[3,4,5]. Typically, conductive hearing loss results from tumor compressing or eroding the ossicular chain, or due to effusion. Most of these signs were found in our patient.

However, due to the slow growing feature of this tumor, the symptoms will often occur when the tumor has already grown. Additionally capable of growing through the tympanic membrane, paragangliomas can be mistaken for an inflammatory tumor. Otorrhagia ca be notice as a significant clinical sign, however, a pulsatile middle ear mass is pathognomonic of a temporal bone paraganglioma[6,5]

The physical examination showed that the finding of purple retrotympanic mass during otoscopy is importan[7], as like as it is found in our patient.

The diagnosis rely on imaging, in particular cerebral MRI, which recovers a typical appearance (Salt and pepper)[8].

The choice of treatment depends on the size and location of the tumour, treatment-related morbidity and mortality, patient age, and patient preferences.

When paragangliomas are localized surgical resection is the treatment of choice[4], however surgery for tumors with intracranial extension is difficult or even inaccessible because of their extension to the neighboring vasculo-nervous structures (the risk of intra cranial hypertension and the impairment of the cranial nerves). However, in some cases radiation therapy is required[9]

However, when surgery is not possible, radiotherapy remains an effective and well-tolerated therapeutic alternative with an excellent local control rate[10].

Conclusion:

Jugular tympanic paraganglioma are benign but rare tumors of the head and neck region. Their risk is mainly represented by the mass effect that they can generate in the ceses with intra cranial extensions. Precise clinical assessment, preoperative imaging and interdisciplinary discussions are the basis of an adequate management of these tumors.

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