SAS Journal of Medicine
Abbreviated Key Title: SAS J Med
ISSN 2454-5112
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Tympano-Jugular Paraganglioma in a Young Patient Treated for Nasal Adenocarcinoma: A Case Report and Review of the Literature

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DOI: 10.36347/sasjm.2023.v09i05.015 | **Received:** 18.04.2023 | **Accepted:** 13.05.2023 | **Published:** 16.05.2023

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Abstract | **Case Report**

Tympano-jugular paraganglioma is a benign tumor with a local aggressive potential and a rare degeneration. The otological signs are prominent and the pulsatile character is pathognomonic. With the use of imaging, the Fish classification can establish the extent and focus the treatment approach. Damage to the cranial nerves predominates among the sequelae.

Keywords: Tympano-jugular paraganglioma, glomus, CT, MRI, Fish classification.

INTRODUCTION

The glomus tumor is a mostly benign proliferation of ectodermal cells of the neural crest [2]. It develops in the carotid paraganglionic tissue, in the jugular foramen, along the vagus nerve or Arnold's nerve and in the tympanic cavity [1, 2].

Despite the good specificity of imaging in this pathology, its occurrence in a previously known neoplastic context should make the diagnosis discussed.

In this paper we report a case of a histologically proven tympano-jugular paraganglioma in a young patient treated for adenocarcinoma of the nasal cavity, reviewing the data in the literature.

OBSERVATION

The patient was a young woman, 24 years old, who had undergone surgery for adenocarcinoma of the nasal cavity. She presented with unilateral pulsatile otalgia that had been present for 4 months.

The clinical examination revealed a patient in good general condition, with a reddish swelling discreetly pulsating in the right external auditory canal. Investigations revealed a conductive hearing loss on the right side with a reddish pulsatile mass in the external auditory canal on otoscopy.

Given the neoplastic context, an MRI of the temporal bone was performed, showing a formation centered on the right temporal bone, roughly oval, with lobulated contours, in T1 isosignal, in T2 heterogeneous hypersignal, intensely and heterogeneously enhanced after injection of gadolinium realizing a salt and pepper aspect (Fig 1), measuring 44 x 37 x 25 mm. This process extends intracranially to the temporal lobe, to the external auditory canal and to the homolateral jugular foramen arriving at the intimate contact of the internal jugular vein.

A CT scan was performed showing a lytic process centered on the temporal bone and extended to the greater wing of the sphenoid and to the acetabular cavity of the temporal bone. The suspected diagnosis on imaging was that of a paraganglioma.

Histological study revealed a myxoid stroma with gaping vessels surrounded by a cellular infiltrate composed of monomorphic cells with eosinophilic cytoplasm and nuclei with fine chromatin. Immunohistochemical study showed that these cells express anti-smooth muscle actin and that CD 34 draws the vascular walls within the proliferation.

The diagnosis retained was a glomus tumor and the patient was referred for surgical management.

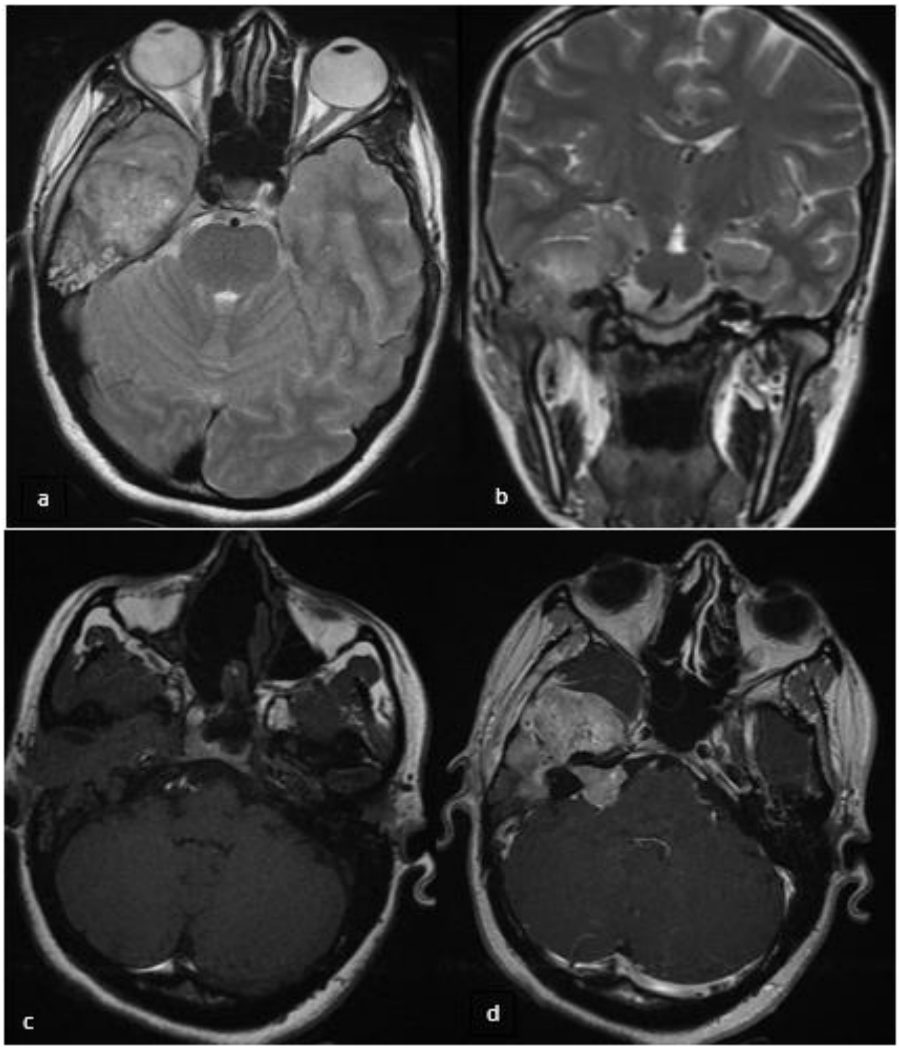

Figure 1: Brain MRI showing the right temporal bone tumor process extended intracranially; a and b : axial and coronal T2 sequence: heterogeneous hypersignal process realizing the salt and pepper sign. c and d: T1 axial sequences before and after gadolinium injection showing heterogeneous enhancement of the tumor.

DISCUSSION

Tympano-jugular glomus tumors are rare and mostly benign tumors, characterized by local aggressiveness and high lytic potential [1]. They occur in elderly subjects between 50 and 60 years of age, with a clear predilection for women (sex ratio: 5/1). The incidence is estimated to be between 1/30000 and 1/100000 and malignant degeneration is possible in 4 to 5% of cases [3].

These tumors originate in the paraganglionic tissue of the vascular or nerve adventitia (internal jugular vein/tympanic branch of the glossopharyngeal nerve: jackobson nerve) [1, 2, 5].

Pathologically, paraganglioma is an encapsulated, hypervascular tumor characterized by the presence of a stroma rich in richly anastomosing sinusoids [2].

The modified Fish classification (Fig 2) differentiates tympanic paragangliomas from tympano-jugular paragangliomas according to the degree of extension; in fact, the latter are characterized by extension outside the tympanic cavity and mastoid with bone lysis [4, 5]. In our case, the tympano-jugular paraganglioma is classified as type D due to the intracranial extension.

The clinical symptomatology is insidious and dominated by the impairment of the auditory function; tinnitus and conductive or mixed hearing loss are the main symptoms. The pulsatile character is pathognomonic [4]. The tympano-jugular forms are associated with invasion of the cranial nerves which may be responsible for facial paralysis (affected nerve VII), dysphagia (affected nerve IX) or Horner's syndrome (affected sympathetic trunk) [3-5].

Otoscopy reveals a pulsatile reddish or bluish retrotympanic mass with a "rising sun" sign [3]. An extension to the external auditory canal is possible.

Due to the hemorrhagic nature of these tumors, histological diagnosis by surgical biopsy is rarely performed and imaging is the cornerstone of the diagnosis in these cases [3, 4]:

- The CT scan shows an isodense tissue mass with irregular and lobulated contours, intensely enhanced after injection of iodinated contrast media. It allows to specify the degree of locoregional extension and bone lysis. Involvement of the jugular foramen suggests a tympano-jugular form.
- On MRI, this tumor is hypo- or isosignal T1, hypersignal T2 heterogeneous by the presence of signal voids corresponding to vessels realizing the "salt and pepper" appearance. Enhancement is intense and heterogeneous after injection of gadolinium. Involvement of the cancellous bone is well detected on MRI by the disappearance of the fatty signal.
- MRI also allows a better analysis of the tumor extension (intra- and extracranial), confirmation of the hypervascular character and a good study of the vascular relationships (permeability of the IJV, internal carotid artery) thanks to the angiographic sequences.
- Angiography shows a hypervascularized mass with rapid venous lavage. It allows a better study of the vascular relationships and identifies the afferent vessels for embolization.

In our case, a surgical biopsy was performed given the context of a known malignant neoplasia in the patient (adenocarcinoma of the nasal cavity) and the diagnosis was confirmed histologically.

The treatment must be chosen in a multidisciplinary consultation and depends on the size, the degree of extension, the age and the general condition of the patient:

- Surgery can be performed but the resection is most often partial in classes C and D; the elements to be taken into account when deciding on surgical management are the degree of encompassment of the internal carotid artery, the intracranial extension, the relationship with the facial nerve and the possibility of preserving the middle ear [4]. In classes A and B, complete surgical excision can be performed.
- External radiotherapy is an alternative to surgical treatment in inoperable cases or in cases of tumor residue after partial resection. Radiosurgery with Gamma-Knife is another therapeutic modality [1].
- Embolization allows a reduction of the tumor volume for surgical treatment.

The evolution may be marked by a recurrence which occurs most frequently at the level of the carotid canal. Post-therapeutic complications (after surgery or radiotherapy) are mainly represented by facial nerve damage and osteomeningeal breaches with CSF leakage.

Tympanomastoid paragangliomas (TMPs)	Class A	Tumors confined to the middle ear	
		A1	Tumor margins clearly visible on otoscopic examination
		A2	Tumor margins not visible on otoscopy. Tumors may extend anteriorly to the Eustachian tube and/or to the posterior mesotympanum
	Class B	Tumors confined to the tympanomastoid cavity without destruction of bone in the infralabyrinthine compartment of the temporal bone	
		B1	Tumors involving the middle ear with extension to the hypotympanum
		B2	Tumors involving the middle ear with extension to the hypotympanum and the mastoid
		B3	Tumors confined to the tympanomastoid compartment with erosion of the carotid canal
Tympanojugular paragangliomas (TJPs)	Class C	Tumors extending beyond the tympanomastoid cavity, destroying bone of the infralabyrinthine and apical compartment of the temporal bone and involving the carotid canal	
		C1	Tumors with limited involvement of the vertical portion of the carotid canal
		C2	Tumors invading the vertical portion of the carotid canal
		C3	Tumors with invasion of the horizontal portion of the carotid canal
		C4	Tumors reaching the anterior foramen lacerum
	Class D	Tumors with intracranial extension	
		De1	Tumors up to 2 cm dural displacement
		De2	Tumors with more than 2 cm dural displacement
		Di1	Tumors up to 2 cm intradural extension
		Di2	Tumors with more than 2 cm intradural extension
		Di3	Tumors with inoperable intradural extension
	Class V	Tumors involving the VA	
		Ve	Tumors involving the extradural VA
		Vi	Tumors involving the intradural VA

Figure 2: Modified Fish classification of paragangliomas of the temporal bone [4]

CONCLUSION

Tympano-jugular paraganglioma is a rare, mostly benign tumor with an aggressive potential. Imaging shows specific signs and is often sufficient for the positive diagnosis and for the orientation of the therapeutic attitude.

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