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Case Report

VERTIGO REVEALING A JUGULAR FORAMEN PARAGANGLIOMA EXTENDED TO THE CEREBELLOPONTINE ANGLE

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ABSTRACT

Jugular foramen paraganglioma (JFP) is a rare, slow-growing and hypervascularized benign tumor. It is generally present in the 5th or 6th decades of life with pulsatile tinnitus and hearing loss. We reported a 71-years-old woman patient with several episodes of Vertigo evolving for few months. Otoscopy revealed a red bulging left sided tympanic membrane. Magnetic resonance imaging (MRI) of the brain showed findings consistent with a large paraganglioma in the left jugular foramen extended to the cerebellopontine angle (CPA). The patient was treated with fractionated stereotactic radiotherapy. JFP should be suspected in a case of vertigo with or without involvement of other cranial nerves.

INTRODUCTION

JFPs are a rare, benign, encapsulated, hypervascular tumors that arise from jugular foramen of temporal bone [1]. They are locally invasive, expanding within the temporal bone via pathways of least resistance, such as air cells, vascular lumens, skull base foramina, and eustachian tube [2]. Their clinical course including various clinical symptoms from lower cranial nerves palsy and pulsatile tinnitus to hearing loss reflects their slow growth and paucity of symptoms, and of ten results in a significant delay in diagnosis [3]. The aim of this work, through a unusual case of JFP to generate awareness and to avoid misinterpretation of this tumor.

Case Report

A 71-years-old woman presented with several episodes of dizziness of a few months duration, that she described as instability and a tendency to fall when walking. She also complained of increasing left hearing loss during 2 years and tinnitus over the last 6 months. Her past medical history was negative from any cerebrovascular accident, migraine headaches, head and neck trauma, or radiation exposure. Otological examination showed bulged left tympanic membrane on its antero-superior surface. Vestibular examination showed scanning speech, vertical nystagmus, and a positive Romberg sign, the patient tending to fall to the right and backwards. Neurological examination noted a paresis of the seventh cranial nerve. Pure tone audiometry showed left sided profound sensorineural deafness. Magnetic resonance imaging (MRI) of the brain showed homogenously enhancing lobulated, extra-axial, altered signal intensity lesion in the region of left jugular foramen, extending into the tympanic cavity, mastoid cells and CPA and compressing the brainstem and the fourth ventricle. The lesion was measuring 34 mm × 21 mm, isointense on T1, iso-to slightly hyperintense on T2 (Fig. 1) weighted images, brilliantly enhancing on contrast (Fig.2), and hyperintense in fluid attenuated inversion recovery sequence.

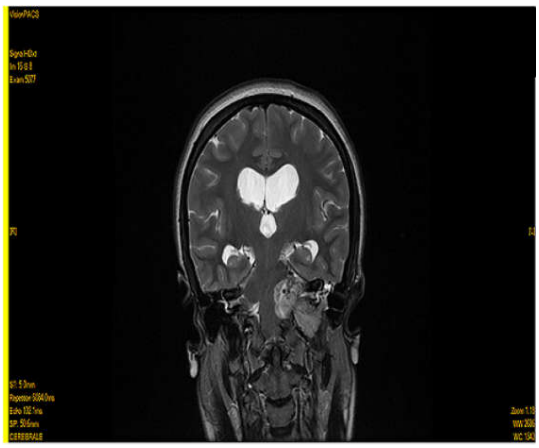

Figure 1 MRI brain with contrast; T2 coronal sequence revealing a mass in the region of left jugular foramen, extending into the tympanic cavity, mastoid cells and cerebellopontine angle.

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Mild restriction was seen on diffusion weighted imaging. The lesion was abutting and partially engulfing the seventh and eighth cranial nerves. These findings appeared to be consistent with jugular foramen paraganglioma. Urine epinephrine, norepinephrine, dopamine and 24-h 5-hydroxyindoleacetic acid and vanillylmandelic acid were all within normal range. The patient was treated with fractionated stereotactic radiotherapy, which she tolerated well. Follow-up MRIs showed partial reduction of the size of the paraganglioma.

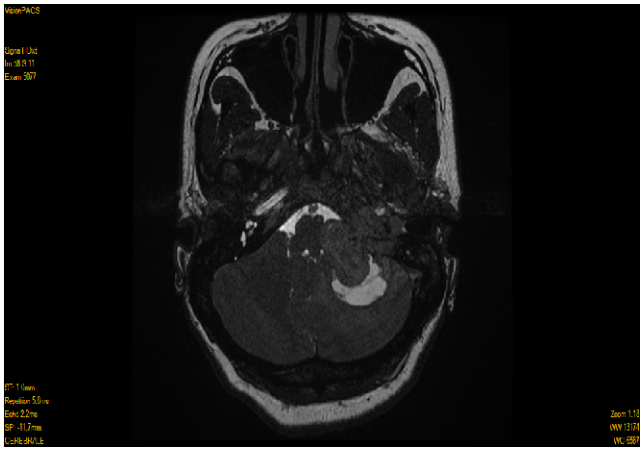

Figure 2 MRI brain with gadolinium contrast; T1 axial sequence demonstrating a lesion at the skull base, at jugular foramen level, mesuring approximately 34 mm × 21 mm extending up to the left CPA cistern and compressing the brainstem

[5]. Our patient presented a left facial paresis, which emphasizes the importance of the neurological assessment of low cranial palsy at the time of the first visit. Emphasis should also be given on the fact that patients without pulsatile tinnitus should not be excluded from the suspicion of JFP. Facial nerve paralysis is known to occur less commonly, but it signals advanced disease and is related with poor facial nerve prognosis outcome. Otoscopic examination can demonstrate a reddish-blue, pulsatile mass but only when the lesion is in contact with the tympanic membrane. Baseline audiometry should always be performed during the initial evaluation to objectively evaluate the degree to which a suspected paraganglioma has affected hearing [6].

Diagnostic imaging is essential to the workup of a suspected jugular paraganglioma to exclude other possible diagnoses and define the extent of disease. High-resolution CT remains the preferred diagnostic modality to visualize the relationship of a suspected tumor to surrounding bone. Erosion around the jugular bulb is considered to be characteristic of a jugular paraganglioma. MRI, with and without contrast, is generally preferred to CT when evaluating the soft tissue component of a tumor. The characteristic appearance of a paraganglioma on contrast-enhanced MRI is the so-called salt-and-pepper pattern most evident on T2-weighted sequences, representing prominent intratumoral flow voids.

Table 1 Classification of jugulotympanic paragangliomas according to Fisch and Mattox.

class	Location and extension of paraganglioma
A	Paragangliomas that arise along the tympanic plexus on promontory
B	Paragangliomas with invasion of the hypotympanon; cortical bone over jugular bulb intact
C1	Paragangliomas with erosion of the carotid foramen
C2	Paragangliomas with destruction of the vertical carotid canal
C3	Paragangliomas with involvement of the horizontal portion of the carotid canal; foramen lacerum intact
C4	Paragangliomas with invasion of the foramen lacerum and cavernous sinus
De ½	Paragangliomas with intracranial but extradural extension;De1/2 according to displacement of the dura (De1 = less than 2 cm, De2 = more than 2 cm)
Di 1/2/3	Paragangliomas with intracranial and intradural extension; Di1/2/3 according to depth of invasion into the posterior cranial fossa (Di1 = less than 2 cm, Di 2 = between 2 and 4 cm, Di3 = more than 4 cm)

DISCUSSION

Paragangliomas are a family of benign, but locally invasive, hypervascular neoplasms that are rare, accounting for only 0.6% of neoplasms of the head and neck region. They are found on the carotid body, the vagus nerve, along the internal jugular vein, or in the tympanic cavity [4].

The JFPs are uncommon, they are known to occur predominantly in the 50-60-years age group and the female-male ratio is reported to be 5:1. The classical evolution of this benign but locally aggressive tumor, is local invasion, follow paths of low resistance towards mastoid cell tracts, vascular channels and eustachian tube, destruction of the petrous bone, invade the CPA [2] such as our case.

The clinical course of JFPs reflects their slow growth and paucity of symptoms, and often results in a significant delay in diagnosis. The most common presenting symptom is pulsatile tinnitus, followed by hearing loss. JFPs can present with symptoms secondary to mass effect on surrounding structures, including vasculature or the lower cranial nerves (VII, IX, X, IX) like facial palsy, dysphagia, hoarseness, aspiration, tongue paralysis, shoulder drop and voice weakness. Neurological examination of lower cranial nerve palsy is crucial since most patients have involvement of at least one lower cranial nerve

Radiologically, paraganglioma needs to be distinguished from acoustic schwannoma and meningioma, which are more common in the CPA. Acoustic schwannoma are round or oval masses in the cerebellopontine cistern that emerge from the internal auditory canal (IAC), widen the porus, and grow posteriorly, and can be heterogeneous due to cystic components. Conversely, meningiomas are usually hemispheric, semilunar, homogenous masses with a broad petrous base to which they are attached and are usually asymmetric to the IAC. If the diagnosis is unclear, a diagnostic angiography can be confirmatory, which shows the specific vascular supply of the paraganglioma[7].

Neurosecretory function in JFPs is rare, but screening for functional tumors remains an important part of tumor management. Functional head and neck tumors are only seen in 1% to 3% of cases and, because of a lack of the enzyme phenylethanolamine-N-methyltransferase, which is not commonly found in extramedullary paragangliomas, norepinephrine secretion predominates.

Tumor staging is an important element of the diagnostic algorithm for GT tumors. Two separate staging systems are commonly referenced: Fisch-Mattox and Glasscock-Jackson. The Fisch-Mattox system incorporates both tympanic and

jugular paraganglioma into one continuum Table.1. The Glasscock-Jackson staging system separates tympanic and jugular tumors [7]. Our case has been classified D1 according to the Fisch-Mattox classification.

Complete surgical resection is the ideal management of most jugular foramen paragangliomas. Surgical excision, though the primary treatment is complicated with per‑operative bleeding because of high vascularity of the tumor. Long‑term control of the disease after surgery for jugular and vagal paranglioma was achieved in 78.2% and 93.3% of patients, respectively. Control of jugular paraganglioma with external beam radiotherapy (EBRT) and stereotaxic radiosurgery was achieved in 89.1% and 93.7% of the cases, respectively. Tumor control failure, major complication rates, and the number of cranial nerve palsies after treatment were significantly higher in surgical than in radiotherapy series [1]. Conventional radiotherapy with fractionated EBRT has been used as a primary, combined, or salvage treatment in patients who cannot undergo surgery because of advanced age or comorbidity, larger, more aggressive, or unresectable tumors, or residual disease [4].

CONCLUSION

JFP is a rare tumor that generally presents, in the 5th or 6th decades of life, with various clinical symptoms from lower cranial nerves palsy and pulsatile tinnitus to hearing loss, depending on the extent of growth. It should be suspected in a case of vertigo with or without involvement of other cranial nerves. Diagnostic imaging is essential to the workup of a suspected JFP to exclude other possible diagnoses and define the extent of disease.

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