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JUGULOTYMPANIC PARAGANGLIOMA WITH MULTIPLE CRANIAL NERVE PALSY : A RARE CASE REPORT

Medicine

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ABSTRACT

Objective: rare disease

Background: Jugulotympanic paragangliomas is a rare tumor that generally presents in the 5th or 6th decades of life with pulsatile tinnitus and hearing loss. In this report, we present an unusual case of jugulotympanic paraganglioma presenting in 48 year old man with multiple cranial nerve palsy and cerebellar ataxia.

Case Report: A 48 year old man presented with diplopia with worsening episodes of dizziness. He also complained of right sided tinnitus and hearing loss along with difficulty in degglution, dysarthia, decreased sensation from right half of face and atropy of right sided tongue muscles. Examination reveals red bulging tympanic membrane right sided along with multiple cranial nerve palsies. CT head suggestive of right sided jugulotympanic paraganglioma which on the MRI shows multiple intracranial and extracranialexpansion with numerous vessels forming AV malformations within the lesion, extending from CP angle down along jugular vein with epicenter in jugular foramen.

Conclusions: Jugulotympanic paragangliomas are rare tumors that usually manifest in middle age with tinnitus and hearing loss. However, clinicians should be aware of unusual presentation of this tumor if presented with multiple cranial nerve palsy and cerebellar ataxia. All the above described clinical features should trigger further investigations with a CT scan or MRI of the brain.

KEYWORDS

Jugulotympanic Paraganglioma ,cp Angle Tumor'

Background

Jugulotympanic paragangliomas is a rare tumor that generally presents in the 5th or 6th decades of life with pulsatile tinnitus and hearing loss. In this report, we present an unusual case of jugulotympanic paragangliomas presenting in 48 year old man with multiple cranial nerve palsy and cerebellar ataxia.

Case Report

A 48 year old man presented with diplopia which is binocular, incomitant and presented in horizontal gaze associated with medial deviation of right eye and gradually progressive loss of vision and decrease field of vision more in temporal side for far objects. Although he had milder form of symptoms for past 10 years, he noticed the apparent worsening of symptoms for last 4 to 5 months. No history of watery secretions, itching, burning sensations, dryness and tear loss in the respective eye and no significant history of trauma and jaw claudication.Patient has several weeks of worsening episodes of "dizziness", which he described as unsteadiness and a tendency to fall without a sense of rotation. These episodes were precipitated by sudden changes in head position but were not preceded by any sensation of ear fullness or ear pain. He also complained of increasing unilateral hearing loss (right side), as well as intermittent tinnitus. He denied any otorrhea,palpitations, weakness, or any recent viral illness. Patient is then complaining of difficulty in deglutition started firstly for large bolus of food, difficulty is more in the initiation of swallowing and food stuck at the pharyngoesophageal junction. In due course of time patient developed water brash symptoms, nasal regurgitation and drooling of saliva from the right side of mouth that is not associated with odynophagia, night sweat anorexia, weightloss. Then patient developed decreased sensations over the right side of face which he describes as not able to detect hot or cold water, along with deviation of face to the left side associated with dysarthria.

Gradually in the last 4 to 5 months patient had developed difficulty in walking in the form of weakness in bilateral lower limb, slowness of movements associated with vertigo and has history of frequent falls, however he has no cognitive difficulty and has no significant changes in behavior. His past medical history was negative for any cerebrovascular accident, migraine headaches, head and neck trauma, or radiation exposure. He denied any history of smoking and alcohol or drug abuse.

On otoscopic examination of the right ear, an erythematous bulging tympanic membrane was noted. Conductive hearing loss was noticed unilaterallyon Rinne test (more on the right side).nystagmus was noted both in horizontal and vertical gaze and Dix-Hallpike test was negative. A bruit was heard at the base of the skull, grade +3 on the right side. A thorough neurological examination produced higher mental function intact. No cognitive and behavior abnormality, wide base gait with swing to both side, no any deformity in spine and cranium is detected, sensations are intact except right sided face, tremors absent, dysmetria present, truncal ataxia present, heel shin test and finger nose test positive dysarthria present.

Cranial nerve examinations revel decrease field of vision and visual acuity in right side more in temporal field. Corneal reflex absent in right side with loss of facial sensations in the same side. 6th cranial nerve palsy in the form of medial deviation of right eye, lower motor neuron lesion of facial nerve (bells phenomenon present). Right sided conductive deafness with sensorineural hearing loss.

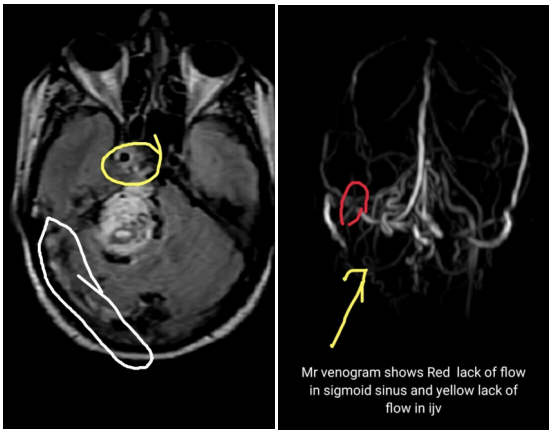
Tongue is deviated to right side with right sided atrophy of muscles with loss of taste sensations right sided. Uvula is deviated to left side with left sided pushed upwards. Gag reflex was absent. Difficulty in shrugging the shoulder of right side s/o rightsided 11 th nerve palsy.

The patient was evaluated by an otorhinolaryngologist, and on otoscopy, the tympanic membrane appeared fluid-filled. Myringotomy with aspiration was attempted with the expectation of a middle ear effusion, but the palpation of a soft doughy mass led to the cessation of the procedure and the concern for either jugulareparaganglioma or tympanic paraganglioma. Computerized tomography (CT) scan of the brain suggestive of a large highly vascular lesion with epicenter in jugular foramen highly likely jugulotympanic paraganglioma showing intracranial and extracranial extension. The lesion is infiltrating right

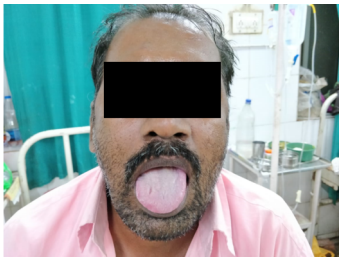
sigmoid sinus, jugular vein and cavernous sinus and encasing right internal carotid artery leading to formation of intracranial arteriovenous malformation MRI of the brain showed a large T1 hypointense and T2 hyperintense lesion showing multiple flow voids signal of vessels, measuring approx. 7.2X 4.1 X 4.8 cm is noted with epicenter most probably in right jugular foramen. Possibility of jugulotympanic paraganglioma is likely. The lesion is causing erosion and expansion of jugular foramen , carotid canal , petrous apex and right half of civus. It is extending outside skull via jugular foramen along with right jugular vein and encasing right styloid process, internal carotid artery, abutting ramus of mandible, infiltrating medial pterygoid muscles and effacing para pharyngeal space.

It is extending into right CP angle and right quadrigeminal cistern, causing mass effect and deviation of brain stem towards left side. Bilateral lateral ventricles and third ventricle are mildly dilated measuring 18mm 9 mm respectively.

The lesion is infiltrating right sigmoid sinus, jugular vein and cavernous sinus and encasing right internal carotid artery leading to formation of arteriovenous malformation with multiple dilated intracranial vessels. A1 SEGMENT OF RIGHT ACA IS NOT VISUALISED SUGGESTIVE OF APLASIA. Rest of the bilateral cerebral hemispheres are normal in attenuation. Right sided otomastoiditis noted.findings consistent with a large paraganglioma on the right sude.Figure 1Figure 2Figure 3Figure 4 Urine epinephrine, norepinephrine, dopamine and 24-h 5-hydroxyindoleacetic acid and vanillylmandelic acid were all within normal range



Facial Deviation to the left side and the right eye is deviated to medial side



Right Sided atrophy of the tongue muscles and the deviation of the tongue to the affected side that is right side

Discussion

Paragangliomas are a family of benign, but locally invasive, hypervascular neoplasms that are rare, accounting for just 0.012% of all tumors [1,2]. They are found on the carotid body, the vagus nerve, along the internal jugular vein, or in the tympanic cavity. Jugulotympanic paragangliomas generally present in the 5th or 6th decades of life with pulsatile tinnitus and hearing loss. In this report, we describe an uncommon presentation of jugulotympanic paragangliomas in the 5th decade of life with multiple cranial nerve palsy and cerebellar ataxia:Vertigo associated with tinnitus and hearing loss suggests Meniere's disease and our patient presented with a similar constellation of symptoms. However, careful examination demonstrated an erythematous bulging tympanic membrane, which prompted further investigations revealing an underlying jugulotympanic paraganglioma.

Jugulotympanic paragangliomas can present with symptoms secondary to mass effect on surrounding structures, including vasculature or the lower cranial nerves [1,3,4] like facial palsy, hearing loss (conductive or sensory), tinnitus, dysphagia, hoarseness, pain, or dizziness. Our patient has facial weakness, dysphagia, diplopia, difficulty in shrugging the right side of shoulder, loss of taste sensations and hemiglossal atrophy were likely from the mass effect of the tumor.

Multiple cranial neuropathies are common entity in neurologic practice. Diagnosis and evaluation of these patients is difficult because of various etiologies and poor outcome. They pass through different structures of cranium and superficial soft tissue, meninges and subarachnoid spaces. Cranial nerves can be affected in their course and present as cranial neuropathyAssymetrical involvement, sequential involvement and simultaneous involvement of distal cranial nerves 5th to 12th indicates a diffuse neoplastic brainstem lesion as well as mass in cerebellopontine angle his hearing loss was unilateral and mixed; which was probably contributed to by the location of the tumor and its expansion causing involvement of multiple cranial nerves.[5, 6]Otoscopic examination of a middle ear paraganglioma can demonstrate a reddish-blue, pulsatile mass medial to the tympanic membrane [7]. Myringotomy is contraindicated in cases of reddish mass behind the tympanic membrane, but in our patient it was performed with the expectation of middle ear effusion with the unanticipated finding of a soft mass behind the tympanic membrane. Catecholamine secretion occurs in only 2% of cases, and the tumor in our case was hormonally inactive, as well. Investigations should include head CT scans to identify the extent of bone destruction and MRI for determining tumor extension; T2-weighted images show "salt and pepper" appearance of the paraganglioma [1,8].[3, 9].

Because of the indolent natural history of these paragangliomas, a period of observation is often appropriate [1, 10]. To be selected for treatment, paragangliomas should be symptomatic or radiologically progressive [11]. With comparable or arguably improved outcomes, and also being 25–30% less expensive than neurosurgery, new irradiation modalities are the first-line treatment for most jugulotympanic paragangliomas [1,12]. The efficacy of radiotherapy is defined not by disappearance of the tumor, but by tumor control (ie., stabilization of symptoms and absence of tumor growth) [12]. The potential late toxicity of radiotherapy suggests that treatment will inevitably evolve towards chemotherapy even though there is no established role for chemotherapy currently [1,12].

Conflict of Interest : NIL

Conclusions

Jugulotympanic paragangliomas are rare tumors that usually manifest in middle age with tinnitus and hearing loss. However, clinicians

should be aware of unusual presentation of this tumor if presented with multiple cranial nerve palsy and cerebellar ataxia. All the above described clinical features should trigger further investigations with a CT scan or MRI of the brain.

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