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A RARE CASE REPORT ON BILATERAL GLOMUS JUGULARE

Radiology

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ABSTRACT

Glomus tumors, also referred to as paragangliomas or chemodectomas, are exceedingly rare neoplasms originating from non-chromaffin chemoreceptor cells along the sympathetic chain. This case report presents a clinical scenario of bilateral glomus jugulare tumors in a 27-year-old male, characterized by progressive hearing loss and vertigo. Advanced imaging modalities, including contrast-enhanced MRI and computed tomography (CT) scans, played a pivotal role in accurately assessing the extent of the lesions, which exhibited well-defined expansile features involving the jugular foramen and accompanying destruction of adjacent structures. Glomus jugulare tumors, ranking as the second most prevalent paragangliomas within the head and neck region, demonstrate familial occurrence with an autosomal dominant inheritance pattern. Furthermore, they pose a diagnostic challenge due to their complex anatomical location, necessitating a multidisciplinary approach for effective management.

KEYWORDS

Glomus tumors, paragangliomas, glomus jugulare, salt and pepper appearance

INTRODUCTION:

Glomus tumors (GT) are also called as paragangliomas or chemodectomas and are rare tumors. These are seen to arise from the non-chromaffin chemoreceptor cells along the sympathetic chain. [1] They may be found in the carotid body, adrenal medulla, and roof of the jugular bulb, along Jacobson and Arnold's nerves, from the jugular fossa to the promontory of the middle ear. Therefore, glomus tumors must be named according to their origin: glomus tympanicum, glomus jugularis, glomus vagale and glomus carotid. Glomus jugulare are second most common head and neck paraganglioma. They arise from the paraganglia associated with the adventitia of the jugular bulb.

Familial occurrence is likely to occur in 10% of patients, with an autosomal dominant inheritance. Multiple tumors are seen in 78-87% of familial paragangliomas, and the incidence of bilateral GT is 32% for familial cases and 4% for non-familial cases. (1)

Case Report:

A 27 year old male, presented with gradual onset of vertigo and right sided hearing loss one year back which subsequently progressed to bilateral hearing loss. On examination, he was of average build, height and well oriented to time, place and person. On otoscopic examination, no significant abnormality was detected. Tuning fork tests and audiometric examination showed hearing loss of mixed nature on either side. Contrast enhanced MRI was performed, which revealed well defined expansile lesions involving bilateral jugular foramen. These lesions showed variable signal intensity of different MRI sequence. On postcontrast fat suppressed T1-weighted sequence, the lesion demonstrated avid contrast enhancement with mixed areas of flow voids consistent with typical 'salt and pepper appearance' of paragangliomas. VII-VIII nerve complexes were normal on either side.

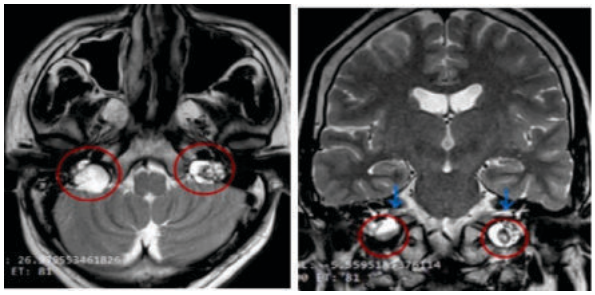

Fig1: T2wighted images

Fig1a*: Saggital view*- T2 mixed signal intensity T2 mixed signal intensity with areas of flow voids (Left> Right), giving it salt and pepper appearance. (red circles)

Fig1b: *Coronal view*- T2 mixed signal intensity T2 mixed signal intensity (red circles). Bilateral VII-VIII nerve complexes are normal (blue arrows).

Subsequently performed plain CT scan reveals expansion and destruction of the jugular foramen on either side with dehiscence of the jugular plate on the right side causing extension of the soft tissue into the middle ear cavity on the right side.

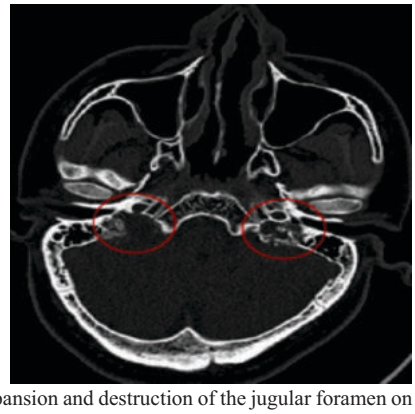

Fig2: Expansion and destruction of the jugular foramen on either side (red circles)

DISCUSSION:

Glomus tumors of the temporal bone, specifically glomus jugulare and glomus tympanicum, manifest within a complex anatomical region intersected by vital neural and vascular structures. These tumors exhibit a range of sizes, spanning from a few millimetres to over ten centimetres. Notably, their growth pattern is gradual, and the majority are classified as benign. Glomus jugulare, the most prevalent type, tends to give rise to substantial masses capable of causing petrous bone destruction and extending into nearby intra/extracranial structures. This particular variant predominantly affects middle-aged women.

Moreover, the familial occurrence of glomus tumors suggests an underlying hereditary factor with an autosomal dominant pattern.

Roughly 25-35% of hereditary cases display multiplicity, whereas the nonhereditary variety accounts for less than 5% (2). Additionally, there have been reports of glomus tumors coexisting with other conditions such as pheochromocytoma, thyroid carcinoma, and neuro fibromatosis [2]. Regarding glomus jugulare tumors, approximately 2-11% of cases exhibit an association with another glomus tumor. While bilateral involvement is possible, the second tumor typically emerges in a distinct location [1].

The optimal imaging techniques employed in the characterization and staging of glomus tumors play a critical role in differentiating them from vascular anomalies, such as a dehiscent high jugular bulb. Computed tomography (CT) is particularly valuable in its ability to precisely delineate the osseous involvement of temporal paragangliomas (PGs). Notably, the expansion and erosion of the jugular foramen are distinctive features observed in jugular PGs. On the other hand, magnetic resonance imaging (MRI) is highly useful in characterizing the lesion itself. T2-weighted MR images often exhibit characteristic small areas of signal void, representing regions of high blood flow within the tumor, resulting in a distinct "salt and pepper" appearance commonly associated with PGs. Gadolinium-enhanced T1-weighted images provide visualization of the tumor's actual vascularity. Magnetic Resonance Angiography (MRA) techniques contribute to determining vascular involvement and the direction of blood flow. [4]

Angiography, as an additional diagnostic tool, reveals glomus tumors as hypervascular masses, featuring enlarged feeding arteries and early venous opacification. While a tumor blush is typically present, it tends to be coarser and less pronounced than that observed in meningiomas. These tumors are primarily supplied by branches of the external carotid artery, most frequently the ascending pharyngeal artery. Other common feeding vessels include the posterior auricular, stylomastoid, and occipital branches. Angiography serves multiple purposes, including diagnosis, assessment of blood supply, and aiding in pre-operative embolization.

Pre-operative embolization of glomus tumors is performed with the primary goals of reducing blood loss during surgery, minimizing the risk of operative complications, and facilitating complete resection to prevent recurrence. Various embolization materials can be utilized, such as polyvinyl alcohol particles (PVA) in sizes ranging from 140 to 250 μm, absorbable embolic materials like sponge particles, and coils. Coils are particularly preferred when anastomoses exist between branches of the external carotid artery and the vertebrobasilar system or internal carotid artery (ICA). It is recommended to allow a delay of at least one to two days between embolization and surgery to allow for the decrease of embolization-related edema, while also avoiding a delay longer than two weeks to prevent recanalization of the feeding vessels. [4].

Glomus jugulare tumors have traditionally been considered surgically inaccessible due to their challenging anatomical location. However, recent advancements in skull base surgery, including refined surgical techniques and instrumentations, have made it possible to safely remove these tumors with reduced morbidity and mortality risks. Despite these advancements, glomus jugulare tumors still present a significant surgical challenge due to their slow growth and often large size at the time of diagnosis.

Given that glomus jugulare tumors originate in the region of the jugular bulb, they can extend into the posterior fossa and involve the lower cranial nerves. Despite notable advancements in surgical and radiation therapy management, determining the optimal treatment approach for these tumors remains a subject of debate. However, most authors concur that maintaining a high index of suspicion for early diagnosis, coupled with prompt and appropriate treatment, can significantly reduce morbidity and mortality in the majority of cases. [3].

CONCLUSIONS:

Bilateral glomus jugulare are very rare tumors and exhibit familial occurrence with autosomal dominant trait. MRI is excellent modality to assess the extent of the glomus jugulare. A comprehensive understanding of the characteristics of glomus tumors, proficient utilization of advanced imaging modalities, and adoption of a multidisciplinary approach are pivotal in effectively managing these rare neoplasms and enhancing patient outcomes.

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