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A case of Glomus jugulare paraganglioma - a rare case report

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Abstract

Glomus jugulare tumors are a rare subset of benign, slow-growing neoplasms originating from neuroectodermal tissues, particularly around the jugular bulb, and can affect the lower cranial nerves. Despite an annual incidence of approximately one case per 1.3 million people, these tumors predominantly impact adults and the elderly, showing a higher prevalence in women and a left-sided preponderance. Symptoms often include tinnitus, hearing loss, and cranial nerve deficits. We present a rare case of a 62-year-old woman with a glomus jugulare paraganglioma involving the vagus and hypoglossal cranial nerves, presenting with a four-year history of hearing loss and pulsatile tinnitus, alongside recent dysphagia and vocal changes. Diagnostic imaging revealed a lobulated, enhancing lesion with a characteristic 'salt and pepper' appearance on MRI, consistent with a glomus jugulare paraganglioma. Despite being advised to undergo surgical resection, the patient opted for radiotherapy, leading to the successful management of the tumor. This case underscores the importance of early diagnosis through effective imaging and highlights the ongoing debate regarding optimal treatment strategies for glomus jugulare tumors. While surgery remains a cornerstone for treatment, particularly in younger patients, radiotherapy, including stereotactic radiosurgery, presents a viable alternative with favorable outcomes and reduced morbidity. This case supports individualized treatment approaches based on patient preferences, tumor characteristics, and potential risks.

Keywords: Glomus jugulare tumor, Paraganglioma, Neuroectodermal tumor, cranial nerve deficits, Pulsatile tinnitus, Hearing loss, Radiotherapy

Introduction

Glomus tumors are benign, slow-growing masses arising from neuroectodermal tissues. In the head and neck, they are classified into two main types, cervical and temporal bone (jugulotympanic) paragangliomas. Glomus tympanicum tumors are more common than glomus jugulare tumors and are the most frequent neoplasms of the middle ear (1). Common symptoms include tinnitus, hearing loss, ear pain, and vertigo (2). In some cases, extensive tumors may affect various cranial nerves (3).

Although rare, with an estimated annual incidence of one case per 1.3 million people, glomus tumors are frequent in the middle ear cavity and temporal bone. They are more common in adults and the elderly, with a significantly higher incidence in women and a left-sided preponderance. Familial cases often present with multiple tumors (4-6). Glomus jugulare tumors originate near the jugular bulb and can extend into the posterior fossa, involving the lower cranial nerves. Despite advancements in surgical and radiation therapy, the optimal treatment approach is still debated. Early diagnosis and appropriate treatment are crucial for reducing morbidity and mortality in most cases.

We present a rare case of glomus jugulare paraganglioma involving the vagus and hypoglossal cranial nerves. Due to a high level of suspicion and effective imaging, an early diagnosis was achieved. Despite the diagnosis, the patient declined surgery and was successfully managed with radiotherapy.

Case Presentation

A 62-year-old woman presented with left ear hard of hearing and pulsatile ringing sound in the left ear for the last 4 years.The patient also had difficulty in swallowing, throat pain, voice change for the last 2 months. No history of dizziness or pain in ear was present.On examination, she was of average build, height and well oriented to time, place and person.

On otoscopic examination, a pulsatile reddish mass was seen behind the intact pars tensa (figure 1), which blanched and stopped pulsating on applying pressure with Siegel's pneumatic speculum. Tuning fork tests and audiometric examination showed hearing loss of mixed nature.On protrusion of the tongue, it deviated to left side suggesting lingual weakness (figure 2).Pure tone audiometry – shows right ear – mild sensorineural hearing loss-32dbHl.Left ear – moderate to severe mixed hearing loss – 82dbHl.On video laryngoscopy – tongue deviated to left side, there is reduced movement of soft palate on left side, there is no movement of left vocal cord, right vocal cord compensatory movement present.

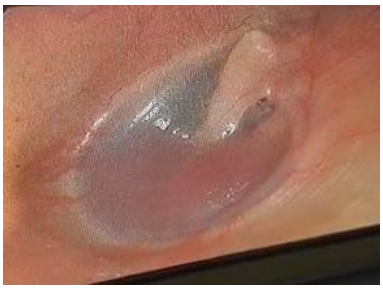

Figure 1: otoscopic examination of left ear showing reddish mass behind the intact pars tensa

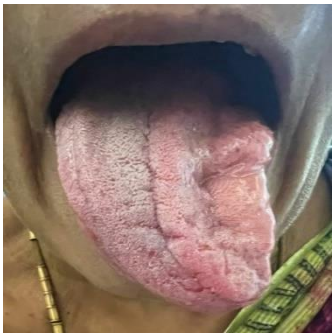

Figure 2: on protrusion of tongue, there is deviation towards left side

On contrast enhanced MRI neck, it was reported as fairly defined, extra-axial, lobulated, heterogenously enhancing lesion along the left cerebello-pontine angle and jugular foramen with internal flow voids and internal linear and punctate non enhancing areas, significant moth-eaten pattern of bony erosions, widened jugular foramen (figure 4).Features represent a neoplastic etiology - Glomus Jugulare paraganglioma.

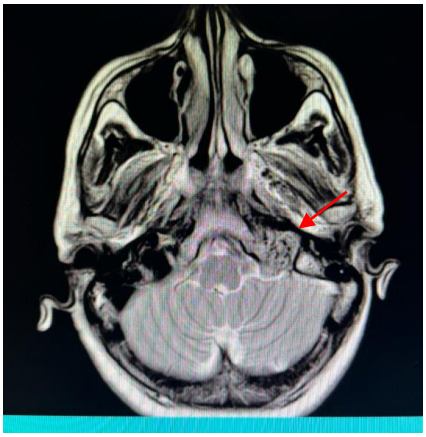

Figure 3: axial section of MRI brain shows salt and pepper appearance of paraganglioma in left cerebellopontine angle

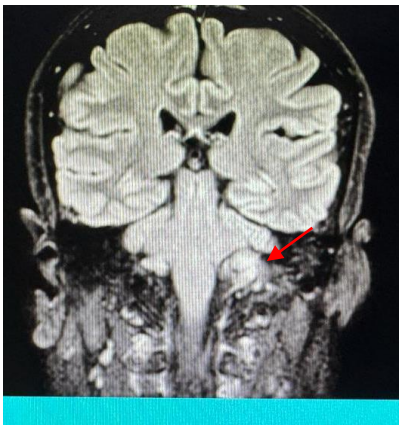

Figure 4: coronal section of MRI brain shows paraganglioma in left

The lesion 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 (figure 3). Twenty-four hours urine vanillylmandelic acid level was within normal range.

CT showed an intra-cranial, extra-axial, infra-tentorial, well-defined hyperdense focus is noted in the left cerebellopontine angle. The location is identified as the left jugular foramen and the right hypoglossal canal, with the epicenter being uncertain. The lesion measures approximately 25 x 25 mm in size and has peripheral smooth to macro-lobulated margins with a central homogeneous appearance. On post-contrast imaging, the lesion is avidly enhancing. The extension and mass effect are observed medially with widening of the left jugular foramen and erosion of the left jugular spine, carotico-jugular spine, and petrous part of the left temporal bone. Posteriorly, there is cortical buckling causing a mild mass effect over the right antero-inferior cerebellum, though likely without invasion, and no evidence of extension to the inner ear. Anteriorly, the carotid canal appears unaffected, and the facial nerve canal remains intact.

Discussion

Potential differential diagnoses for glomus jugulare include jugular Schwannoma, neurofibroma, meningioma, metastasis, and primitive neuroectodermal tumor. Observation is a viable treatment option, as 65% of tumors remain stable or regress in size (7). Approximately 40% of tumors grow at an average rate of 0.9 mm per year (8). For patients diagnosed with glomus jugulare who opt for observation, close follow-up with serial brain MRI, both with and without intravenous contrast, is essential to monitor the disease's progression.

Various therapeutic options aim to improve local control and minimize treatment-related morbidity, but managing glomus tumors remains challenging (9). There is still considerable debate regarding the optimal treatment. Historically, treatment involved surgical resection, external beam radiotherapy (EBRT), or a combination of both (10). However, these methods carry significant morbidity risks, making stereotactic radiosurgery (SRS) an increasingly popular option (10). Systematic reviews indicate that EBRT and SRS offer outcomes comparable to surgical intervention for jugular paragangliomas (11). Treatment decisions should be individualized for each patient.

For young, healthy patients with functional cranial nerve deficits, surgical removal is the preferred treatment. Preoperative embolization is typically performed 24 to 72 hours before surgery (12). Complete resection is possible in about 80% of patients but may lead to debilitating cranial neuropathy. Postoperative cranial nerve injuries occur in 60% of patients, involving nerves IX, X, XI, and XII. Subtotal resection is increasingly used to minimize morbidity and alleviate symptoms associated with the disease (8).

Subtotal resection followed by radiosurgery for residual tumors yields better outcomes with lower morbidity and mortality (13). Endoscopy, often used for posterior fossa extensions, allows for smaller incisions and tailored trajectories. Advances in technology have enabled nerve monitoring techniques to prevent lower cranial nerve damage during surgery.

Radiation can be used for bilateral glomus jugulare tumors and as an adjunct to limited surgical approaches with subtotal resection (9). Treatment modalities include standard fractionated radiotherapy and stereotactic radiosurgery. Standard fractionated radiotherapy requires multiple sessions, while radiosurgery provides focused radiation in a single session, sparing vulnerable structures inside the tympanic bone (9). Radiosurgery can be an adjunct to limited surgery or a primary treatment for poor surgical candidates or patients with bilateral disease, offering up to 90% control rates (9, 14). Radiosurgery alone provides tumor control

in 92% of patients, symptom control in 93%, with an 8% complication rate (15, 16). For local control rates of ≥90%, a median marginal dose of 15 Gy (range, 12-30 Gy) is recommended (17).

Complete obliteration of glomus jugulare tumors through embolization is challenging due to the potential for revascularization and limited effectiveness in alleviating symptoms (18). Endovascular embolization as a sole treatment is considered palliative (9). Onyx embolization can be used for palliative management of otorrhagia in patients with unresectable tumors (19). Preoperative embolization can reduce surgery duration and operative blood loss (9).

Conclusion

Glomus jugulare tumors, which originate near the jugular bulb, can sometimes extend into the posterior fossa and affect the lower cranial nerves. Despite substantial advancements in managing this tumor through surgery and radiation therapy, determining the optimal treatment remains a subject of ongoing debate. However, most experts agree that early diagnosis, coupled with timely and appropriate treatment, can significantly reduce morbidity and mortality in most cases.

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