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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 . These tumors primarily affect adults and the elderly, with a left-sided preponderance and a greater prevalence in women, despite a yearly incidence of about one case per 1.3 million persons. Tinnitus, hearing loss, and cranial nerve impairments are common symptoms .In this uncommon case, a 62-year-old woman with a glomus jugulare paraganglioma affecting the vagus and hypoglossal cranial nerves presents with recent dysphagia and vocal abnormalities, as well as a four-year history of hearing loss and pulsatile tinnitus. Diagnostic imaging showed a lobulated, enhancing lesion that was compatible with a glomus jugulare paraganglioma. The lesion had a distinctive "salt and pepper" look on MRI. The patient chose to receive radiotherapy instead of surgical resection, which resulted in the tumor being successfully managed .This case shows the ongoing discussion about the best ways to treat glomus jugulare tumors and emphasizes the significance of early diagnosis by efficient imaging. Surgery is still the mainstay of treatment, especially for younger patients, but radiotherapy—including stereotactic radiosurgery—offers a good substitute with better results and lower morbidity. Individualized treatment plans based on patient choices, tumor characteristics, and possible dangers are supported by this instance.

Keywords

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

INTRODUCTION

Benign, slowly expanding masses that originate from neuroectodermal tissues are called glomus tumors. There are two primary forms of paragangliomas in the head and neck: cervical and temporal bone (jugulotympanic) paragangliomas. The most common middle ear neoplasms are glomus tympanicum tumors, which are more common than glomus jugulare tumors (1). Tinnitus, vertigo, ear discomfort, and hearing loss are typical symptoms (2). Numerous cranial nerves may occasionally be impacted by large tumors (3).

Glomus tumors are common in the temporal bone and middle ear cavity, although being uncommon—an estimated one case per 1.3 million people occurs each year. They are more prevalent in adults and the elderly, with a left-sided preponderance and a noticeably higher prevalence in women. Multiple tumors are common in familial instances (4-6). Tumors of the glomus jugulare, which affect the lower cranial nerves, start close to the

jugular bulb and may spread into the posterior fossa. The best course of treatment is still up for discussion, despite improvements in radiation and surgery. In the majority of cases, lowering morbidity and mortality requires early diagnosis and suitable treatment.

We report a rare instance of glomus jugulare paraganglioma that affects the hypoglossal and vagus cranial nerves. Effective imaging and a high degree of suspicion led to an early diagnosis. The patient refused surgery in spite of the diagnosis, and radiotherapy was an effective treatment.

CASE PRESENTATION

A sixty-two-year-old woman came with complaints of left ear hard of hearing and a pulsating ringing sound that had been there for four years. The patient has also experienced voice change , throat pain, and difficulty in swallowing for the past two months. There was no history of ear pain or vertigo. According to the examination, she was of average height and build, and she was aware of time, place, and people.

An otoscopic examination revealed a pulsating reddish mass behind the intact pars tensa (figure 1), which blanched and ceased pulsing when Siegel's pneumatic speculum was applied. Mixed-type hearing loss was revealed by audiometric testing and tuning fork tests.The tongue deviated to the left when protruding, indicating lingual weakness (figure 2). The right ear has a mild sensorineural hearing loss of 32 dBHl, according to pure tone audiometry. Moderate to severe mixed hearing loss (82dbHl) in the left ear. Video laryngoscopy shows a left-sided tongue deviance, decreased soft palate movement on the left side, no left vocal cord movement, and compensatory movement in the right vocal cord.

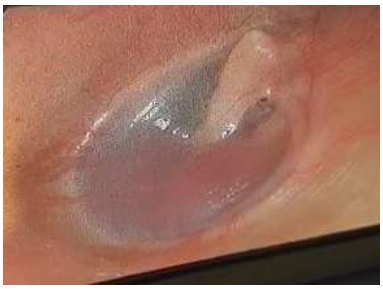

Figure 1: otoscopic examination of left ear showing reddish mass behind the intact pars tensa

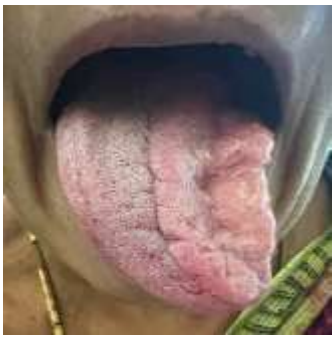

Figure 2: on protrusion of tongue, there is deviation towards left side

A fairly well-defined, extra-axial, lobulated, heterogeneously enhancing lesion along the left cerebello-pontine angle and jugular foramen was observed on the contrast-enhanced MRI neck. It was described as having a significant moth-eaten pattern of bony erosions, a widened jugular foramen, and internal flow voids as well as internal linear and punctate non-enhancing areas (figure 4). Features indicate Glomus Jugulare paraganglioma, a neoplastic etiology.

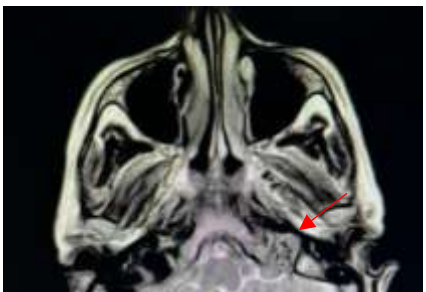

Figure 3: axial section of MRI brain shows salt and pepper appearance of paraganglioma in left cerebellopontine angle

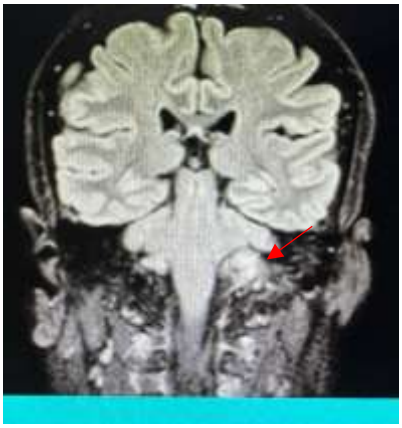

Figure 4: coronal section of MRI brain shows paraganglioma in left

The lesion displayed varying MRI sequence signal intensities. The lesion showed avid contrast enhancement on the postcontrast fat-suppressed T1-weighted sequence, with mixed areas of flow voids that were typical of paragangliomas' "salt and pepper appearance" (figure 3). The level of vanillylmandelic acid in the urine after 24 hours was within the normal range.

CT revealed a well-defined hyperdense focus in the left cerebellopontine angle that was intracranial, extra-axial, and infra-tentorial. The epicenter is unknown, but the location is determined to be the right hypoglossal canal and the left jugular foramen. The lesion is roughly 25 x 25 mm in size, with a homogeneous appearance in the center and smooth to macrolobulated margins on the periphery. The lesion is actively growing on post-contrast imaging. The left jugular foramen widens, the left jugular spine, the carotico-jugular spine, and the petrous portion of the left temporal bone erode, and the extension and mass effect is seen medially. There is no indication of extension to the inner ear, and cortical buckling posteriorly causes a slight mass effect over the right antero-inferior cerebellum, most likely without invasion. The facial nerve canal is still intact, and the carotid canal doesn't seem to be impacted anteriorly.

DISCUSSION

Jugular Schwannoma, neurofibroma, meningioma, metastasis, and primitive neuroectodermal tumor are possible differential diagnosis for glomus jugulare. Given that 65% of tumors either stay stable or shrink in size, observation is a good course of treatment (7). The average annual growth rate of 40% of tumors is 0.9 mm (8). Close follow-up with serial brain MRI, both with and without intravenous contrast, is necessary to track the disease's course in patients with glomus jugulare who choose observation.

Glomus tumor management is still difficult, despite a number of therapy strategies that try to enhance local control and reduce treatment-related morbidity (9). The best course of treatment is still hotly contested. External beam radiation therapy (EBRT), surgical excision, or a combination of the two have been used as treatment in the past (10) .But because of the high morbidity risks associated with these techniques, stereotactic radiosurgery (SRS) is becoming a more and more preferred choice (10). According to systematic evaluations, for jugular paragangliomas, EBRT and SRS provide results that are on par with surgery (11). Every patient should have a unique treatment plan.

The recommended course of treatment for young, healthy patients with functional cranial nerve impairments is surgical removal. Usually, preoperative embolization takes place 24 to 72 hours before to surgery (12). In over 80% of cases, complete resection is feasible; nevertheless, it may result in crippling cranial neuropathy. Nerves

IX, X, XI, and XII are affected by postoperative cranial nerve injuries, which affect 60% of patients. Subtotal resection is being used more often to reduce morbidity and ameliorate disease-related symptoms (8).

Partial resection followed by radiosurgery for remaining tumors results in improved outcomes with reduced morbidity and mortality (13). Endoscopy, frequently utilized for posterior fossa extensions, facilitates smaller incisions and customized trajectories. Technological advancements have introduced nerve monitoring methods to prevent damage to lower cranial nerves during the procedure.

Radiation therapy can be applied to bilateral glomus jugulare tumors and as a complementary treatment to limited surgical techniques involving partial resection (9). Treatment options consist of conventional fractionated radiotherapy and stereotactic radiosurgery. Conventional fractionated radiotherapy involves multiple sessions, whereas radiosurgery delivers targeted radiation in a single session, protecting delicate structures within the tympanic bone (9). Radiosurgery may serve as a supplementary option to limited surgery or act as a primary treatment for patients who are not suitable for surgery or have bilateral conditions, achieving control rates of up to 90% (9, 14). Using radiosurgery alone results in tumor control for 92% of patients and alleviates symptoms for 93%, although there is an 8% rate of complications (15, 16). To achieve local control rates of at least 90%, a median marginal dose of 15 Gy (with a range of 12-30 Gy) is suggested (17).

Complete removal of glomus jugulare tumors via embolization is difficult due to the risk of revascularization and its limited effectiveness in symptom relief (18). Endovascular embolization used as the only method of treatment is regarded as palliative (9). Onyx embolization can be utilized for the palliative management of otorrhagia in patients with tumors that cannot be surgically removed (19).Preoperative embolization can reduce surgery duration and operative blood loss (9).

CONCLUSION

Glomus jugulare tumors, which develop close to the jugular bulb, occasionally extend into the posterior fossa and impact the lower cranial nerves. Although there have been significant improvements in managing this tumor via surgical and radiation methods, the best treatment approach is still a matter of discussion. Nevertheless, the majority of specialists concur that early detection, combined with prompt and suitable treatment, can greatly decrease morbidity and mortality in most instances.

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