Glomus Jugulare: A Case Of Secretory Glomus Jugulare With Review Of Literature

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Abstract

We report a case of secretory glomus jugulare tumor in a 65 year old female who presented with hearing impairment, pulsatile tinnitus, from 3 years and difficulty in swallowing, hoarseness of voice and palpitation from last six months. Detailed history and examination with aid of investigations it turned out secretory glomus jugulare. Due to patients infirmity, she was treated with radiations and is doing well from last 4 years.

Introduction

Glomus jugulare tumors are rare, slow growing, hypervascular tumors that arise within the jugular foramen of the temporal bone. Glomus jugulare tumors originate from the chief cells of the paraganglia, or glomus bodies, located within the wall (adventitia) of the jugular bulb, and can be associated with either the auricular branch of the vagus nerve (Arnold nerve) or the tympanic branch of the glossopharyngeal nerve (Jacobson nerve). Estimated incidence is 1 per 1.3 million population of which 1-5% are malignant. The benign tumors are locally aggressive. Growth rate is about 0.8mm per year. Glomus secreting catecholamines occur symptomatically in about 1-3% of cases.

Case Report(s)

A 65 year old lady, reported with chief complaints of impairment of hearing right ear and pulsatile tinnitus from last three years with difficulty in swallowing and hoarseness of voice, palpitation from last six months. There was no history of vertigo or facial weakness. Past history revealed hypertension 8 years on beta-blockers. Physical examination of the patient revealed a conscious, cooperative well built averagely nourished female. Vitals include a heart rate of 76 beats /minute, respiratory rate of 12 per minute, afebrile, with normal gait. However, pulse was irregular and BP 160/68 mmHg. Carotid pulsations on right side was very prominent with bruit. Otoscopy showed a pulsatile reddish mass behind intact ear drum involving part of anterio-inferior and posterior-inferior quadrant of pars-tensa, on application of pressure to the external auditory canal with the help of pneumatic otoscope, there is cessation of pulsation and blunching of mass. There was also tongue fasciculation with atrophy of right side of the tongue and on protrusion of tongue, tongue deviated to right side. However decreased palatal movements on right side with deviation of uvulae to the left side and gag reflex was absent in posterior pharyngeal wall on the right side. There was also right vocal cord palsy and weak shrugging of shoulder on right side. Tuning fork tests and pure tone audiometry showed mixed hearing loss. Contrast enhanced CT scan of head and neck showed an enhancing mass lesion in right jugular fossa extending to right petrous carotid canal with erosion and destruction of petrous part of temporal bone without intracranial extension. MRA and MRV showed evidence of expansion of right jugular fossa with an ill defined lobulated 2x2.5cm speckled mass lesion with mixed signal intensity and absent flow within right jugular vein, non-visualized right sigmoid sinus. Rest of the dural venous sinuses are normal. Vanillyl-mandelic acid (VMA) in 24 hour urine by high per formance liquid chromatography was 6.2mg/g creatinine (normal range 1.6-4.2). On this basis secretory glomus jugulare was diagnosed. As per the classification devised by Oldring and Fish, the tumor was type C. As tumor was extending beyond petrous apex, affecting lower cranial nerves, patient was 65 years and medically infirm, patient refused surgery, and we treat the patient with radiotherapy.

Discussion

Glomus jugulare tumor, once thought to be one of the most difficult surgically unapproachable ones, are now becoming safely manageable with reasonable morbidity and mortality rates. This recent achievement has been accomplished by the extraordinary efforts put forth in the understanding of the microsurgical techniques and instrumentations and by the most exciting and promising innovations in cranial base surgery. Because of the insidious onset of symptoms,
affected sites that are much less common. After 1953, Guild reported in 2002 on 1,9,10, 14, 15 described a similar modification as the most common tumor of the temporal bone. The female-to-male ratio is 3:6:1. Glomus jugulare tumors have also been noted to be more common on the left side, especially in females. Most tumors occur in patients aged 40-70 years, but cases have been reported in patients as young as 6 months and as old as 88 years. Multicentric tumors are found in 3-10% of sporadic cases and in 25-50% of familial cases. 1,8,10.

In about 2-4% of cases, the first or leading symptoms are hypertension and tachycardia (pheochromocytomalike symptoms) produced by catecholamines, norepinephrine, or dopamine excreted by the tumor. Also, somatostatin, vasoactive intestinal polypeptide (VIP), calcitonin, and neuron-specific enolase may be produced by the tumor. Other related symptoms include headache, perspiration, pallor, and nausea. Otoscopic examination reveals a characteristic, pulsatile, reddish-blue tumor behind the tympanic membrane that is often the beginning of more extensive findings (ie, the tip of the iceberg). Audiologic examination reveals mixed conductive and sensorineural hearing loss. The sensorineural component tends to be more significant with larger tumors. Plain skull radiography may show enlargement of the lateral jugular foramen and fossa. Axial and coronal computed tomography (CT) scanning with thin sections are superior at demonstrating the extent of bone destruction. Magnetic resonance imaging (MRI) with gadolinium-diethylene triamine pentaacetic acid (DTPA) contrast is best for delineating tumor limits. Glomus tumors on T1- and T2-weighted MRI have characteristic soft tissue mixed intensity with intermixed high-intensity signals and signal voids (ie, salt and pepper appearance) representing fast flowing blood. A combination of CT scanning and contrast MRI is the imaging regimen of choice for glomus jugulare tumors. Unless carotid arteriography is necessary for preoperative evaluation and/or embolization, noninvasive techniques are preferred; however, for large tumors involving the internal carotid artery (ICA), preoperative carotid arteriography with cross-compression or trial balloon occlusion is recommended. The venous drainage systems also need to be carefully studied before sinus occlusion is carried out during surgical resection. For tumors with large intracranial extension, vertebral arteriography is advised to exclude arterial feeders from the posterior circulation. 1,2,3,4,7,8, 11 A variety of functional imaging studies for neurosecretory tumors are also available. Radioactive labeled meta-iodobenzyl guanidine (MIBG) is taken up by the neurosecretory cells because of structural similarity to noradrenaline. Therefore 123I-MIBG scan can help in detecting secretory paragangliomas. The sites picked up by the scan can be targeted with 131 I-MIBG loaded with substantial radioactivity in malignant cases. 12

Medical Therapy: Some cases require no treatment. Often, glomus jugulare tumors are diagnosed within the sixth or seventh decade of life and can be followed by imaging only and may not need surgical intervention. Medical therapy may be indicated in some cases. Alpha-blockers and beta-blockers are useful for tumors secreting catecholamines. They are usually administered for 2-3 weeks before embolization and/or surgery to avoid potentially lethal blood pressure lability and arrhythmias. Successful treatment of pulmonary metastases with etoposide (VP-16) and cisplatin has been described.

Surgical Therapy: After 1953, Guild significantly enhanced our understanding of this entity it was in 1987, Al-Mefty, et al, 13 described a combined infratemporal and posterior fossa approach for the removal of giant glomus jugulare tumors that have a large intracranial component. This approach provided access to tumors previously thought to be inoperable and obviated the need for multiple stages of surgery. In 1989, Bordi, et al, 14 described a similar modification of the infratemporal and lateral approach, which they called the "single-staged posterolateral combined otoneurosurgical approach." In 1994, Patel, et al, 15 published a report on a series of 12 patients with extensive glomus jugulare tumors in which the subtemporal–infratemporal, retrosigmoid, and/or extremelateral transcnoydar approaches were used in combination with the usual transtemporal–infratemporal approaches described previously. Al-Mefty and Teixeira 16 reported in 2002 on the management of tumors of the glomus jugulare that they termed "complex." To be classified as complex, one or more of the following criteria had to be met: giant size, multiple paragangliomas, malignancy, catecholamine secretion, association with other lesions, previous treatment with adverse outcome, radiation therapy, or adverse effects from embolization.
Similarly to Patel and colleagues, they stressed the modification of existing approaches to achieve adequate exposure of the lesion. A major advancement in the surgical treatment of glomus jugulare tumors occurred with the development of preoperative superselective embolization. After it was introduced by Hilal and Michelsen\textsuperscript{17} and Brismar and Cronqvist\textsuperscript{18} in separate publications, in 1979 Simpson, et al.\textsuperscript{19} reported the use of preoperative embolization in glomus jugulare tumors in an effort to reduce intraoperative blood loss. Murphy and Brackmann\textsuperscript{20} substantiated the use of preoperative embolization in a 1989 report in which 35 patients were analyzed. They concluded that there was a significant reduction in both intraoperative blood loss and operating time. In addition, embolization led to a higher rate of complete resection. Nevertheless, there did not appear to be a reduction in the risk of injury to the lower cranial nerves. As for the morbidity associated with the embolization procedure, the current state of technology and expertise in interventional radiology has significantly reduced the incidence of stroke and cranial nerve injury experienced during the early years of its application. Surgical approaches are classified into

(i) **Posterior (through posterior cranial fossa):** Sub-occipital retrosigmoid trans-condylar, supracondylar approaches.

(ii) **Anterior:** Pre-auricular subtemporal, infratemporal approaches.

(iii) **Lateral approach:** Juxtacondylar and lateral skull base approaches.

**Contraindication:** Because this tumor is rare and may present with various symptoms, surgery may be contraindicated for various reasons, including age and general physical condition. Surgical resection of the glomus tumor is relatively simple and complication free for type I tumors. Large tumors that affect the lower cranial nerves and extend beyond the petrous apex carry a significant risk of postoperative complications, especially in older patients.

**Radiation Therapy:** The treatment of glomus jugulare tumors with radiation therapy remains controversial. To date, there is no conclusive data establishing radiation as the optimal primary treatment for all glomus jugulare neoplasms. Spector, et al.\textsuperscript{9} revealed in 1973 that radiation therapy had relatively little effect on the tumor cells, with the most dramatic changes consisting of a marked increase in the stromal fibrous connective tissue. Several other authors have reported that the primary effect of radiation therapy is a radiation-induced vascular injury.\textsuperscript{11} Nonetheless, glomus tumors treated primarily with radiation have been reported in multiple clinical studies describing excellent tumor control with only rare cases of tumor progression.\textsuperscript{21} Two points must be made concerning the conclusions reached in these studies. First, an overwhelming number of patients were followed up for less than 5 years, and it is a well-known fact that recurrent tumors can arise up to 25 years after the initial treatment.\textsuperscript{22} The second point is that a majority of patients treated with radiation had no change in the size of their tumor. Regardless of the shortcomings of radiation therapy, surgical treatment involves the risks associated with induction of general anesthesia and carries the potential for cranial nerve injury. These problems are not encountered when fractionated radiation is used. For patients who are medically unfit for surgery, of an advanced age, or at significant risk for cranial nerve injury, radiation therapy may be of significant benefit. In several recent studies the use of gamma knife surgery in the treatment of glomus tumors has been investigated. In 1997, Foote, et al.\textsuperscript{23} published the first report as a preliminary study. The goal of their study was to evaluate the immediate, acute, and chronic toxicity and the efficacy of stereotactic radiosurgery in patients with unresectable or subtotally resected glomus tumors. No acute or chronic toxicity was demonstrated, and eight of nine tumors remained stable in size at a median clinical follow-up duration of 20 months. In 1999, Eustacchio, et al.\textsuperscript{24} reported on 10 patients, with radiosurgery being used as the primary treatment in seven of them. The median follow-up duration was 37.6 months, with 60% of patients exhibiting no change in tumor size, and with the remainder showing decreased tumor volumes. In a series published by Jordan, et al, in 2000\textsuperscript{25}, eight patients who were deemed unsuitable for surgery were treated with stereotactic radiosurgery. One patient experienced intractable vertigo requiring hospitalization. The mean follow-up duration was 27 months, and no patient had an increase in the size of the tumor. Saringer, et al.\textsuperscript{26} reported on 13 patients in 2001, with none showing evidence of tumor growth. Two patients experienced radiation-induced cranial neuropathies, both of which were transient. In a follow-up study to their 1996 series, in 2002 Foote, et al.\textsuperscript{27} reported on 25 patients, 16 of whom were added after the first study. There was one episode of vertigo in their series, similar to the case reported by Jordan, et al.\textsuperscript{28} The median follow-up duration was 35 months, and no tumors exhibited growth during that time. As with most of the series involving fractionated radiation therapy, the follow-up period for these studies is short. We are awaiting future reports with longer follow-up times to demonstrate the efficacy of stereotactic...
radiation therapy in the management of glomus jugulare tumors. Nonetheless, the results are promising.

Conclusion

High index of suspicion, with early diagnosis and appropriate treatment to secretory glomus jugulare, can reduce the morbidity and mortality to even infirm elderly women as has been seen in our case.

Abbreviations(s)

Nil

Authors contribution(s)

Equal contribution by all

References

Illustrations

Illustration 1

Patient having right sided tongue palsy with visible atrophy
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