Contemporary Management of Jugular Paragangliomas

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KEYWORDS

• Jugular paraganglioma • Cranial nerves • Glomus tumor • Jugular foramen • Carotid artery

KEY POINTS

- Jugular paragangliomas are the most common tumors of the jugular foramen.
- The management of jugular paragangliomas is challenging because of their close proximity to cranial nerves (CN) and the internal carotid artery.
- Surgery, radiation, and observation are all viable management options and should be individualized to the patient.
- At the authors' center, there has been a paradigm shift toward conservatism in selected cases in order to minimize morbidity.

INTRODUCTION

Jugular paragangliomas (JPs) are the most common primary neoplasms of the jugular foramen, arising from the paraganglion cells within the adventitia of the jugular bulb. They are slow-growing, highly vascularized tumors that are usually diagnosed during the fourth to fifth decades of life, affecting women 3 times more frequently than men. Although considered histologically benign, the management of jugular paragangliomas is challenging because of their infiltrative nature and close proximity to the facial nerve and lower cranial nerves (CN), carotid canal, posterior fossa meninges, and otic capsule.[1–4] Historically, gross total microsurgical resection was considered the

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Abbreviations

CN Cranial nerves
GJT Glomus jugulare tumor
JP Jugular paragangliomas

treatment of choice, offering complete eradication of disease; however, this strategy may cause significant morbidity, even in the hands of experienced surgeons.[5]

In an effort to explore less invasive treatment methods, stereotactic radiosurgery began gaining popularity in the early 1990s, and today has become the primary treatment modality of choice for many centers. The primary benefit of radiation therapy is a lower risk of up-front cranial neuropathy compared with gross total resection; however, tumor control and length of follow-up in these studies are variable.[6,7]

More recently, observation has been considered for select patients such as those with small tumors and few attributable symptoms, those with multicentric disease and contralateral lower cranial neuropathy, or elderly and infirm patients without brainstem compression. The data concerning observation for JP are scarce, and few centers have looked into the clinical course of untreated JP.[8,9] Such data are needed in order to compare against outcomes with radiation therapy. For example, if it was demonstrated that a large number of tumors do not grow for extended periods of observation, it could be argued that radiation therapy should be reserved until there is definitive evidence of growth.

The Otology Group of Vanderbilt has over 40 years of experience with JP. In the authors' practice, most tumors are managed with microsurgery; however, over the last decade, the authors' group has adopted a less aggressive approach in select patients in order to minimize cranial nerve morbidity. In this article, the authors report their experience managing JP, highlighting the paradigm shift in treatment at the authors' center.

DISEASE PRESENTATION

Pulsatile tinnitus is the most common presenting symptom in patients with JP, followed by hearing decline.[10] Hearing loss is usually conductive in nature but can be sensorineural or mixed.[11] Lower cranial neuropathies resulting in dysphagia, hoarseness, shoulder weakness, and tongue hemiparesis are less common and are usually seen with larger tumors that extend through the medial wall of the jugular bulb. Headache and vomiting are usually late signs associated with increased intracranial pressure caused by brainstem compression and fourth ventricle effacement.[12]

A pulsatile red middle ear mass behind an intact tympanic membrane is the most common finding on physical examination (**Fig. 1**). By definition, a glomus jugulare extends from the jugular bulb and hypotympanum into the middle ear space. Therefore, the middle ear component only represents the tip of the iceberg. Although not universally present, increased canal and tympanic membrane vascularity surrounding the inferiorly based middle red ear mass may result in the characteristic, rising sun appearance. Less commonly, the tympanic portion of the tumor may erupt into the ear canal, resulting in bloody otorrhea.

In contrast to visceral paragangliomas, head and neck paragangliomas are rarely (<4%) secretory.[13] Patients reporting a history of palpitations, sweats, flushing, syncope, hypertension, and headaches should be screened for serum and urine catecholamine levels. If elevated catecholamine levels are found, the patient should undergo further imaging to rule out pheochromocytoma or multicentric paraganglioma disease.