Intracranial Hypertension due to Glomus Jugulare and Contralateral Sigmoid Sinus Stenosis

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ABSTRACT

Educational Objective: At the conclusion of this presentation, the participants should be able to recognize the diagnostic conundrum of unilateral glomus jugulare presenting with intracranial hypertension.

Objective: To present a case of unilateral paraganglioma with simultaneous contralateral sigmoid sinus stenosis causing bilateral venous outflow obstruction leading to intracranial hypertension.

Study Design: Retrospective case report.

Setting: Tertiary referral center.

Methods: The clinical course, radiologic imaging, and procedural details of a single patient were reviewed. A literature search was conducted (PubMed, 1950 to July 2016) for paraganglioma and dural venous sinus stenosis.

Results: A 58-year-old female presented with a right-sided pulsatile hearing loss, pulsatile tinnitus, and aural fullness. Otoscopy was notable for a red, pulsatile mass in the inferior quadrant of the right tympanic membrane. Imaging revealed a right-sided glomus jugulare tumor causing right jugular bulb and sinusoid sinus obstruction. A complete workup including laboratory testing for a secreting tumor was performed. Normal metanephrines demonstrated a non-secreting tumor and the patient subsequently underwent CyberKnife stereotactic radiosurgery for treatment. She began experiencing progressive headaches and visual changes which were not explained by a nonsecreting paraganglioma. Further workup including angiography identified intracranial hypertension secondary to idiopathic left-sided proximal sigmoid sinus stenosis, which, along with her right-sided paraganglioma, resulted in bilateral venous outflow obstruction. Endovascular stenting of the left-sided sigmoid sinus stenosis led to improvement in venous pressure gradients as well as complete resolution of headaches and normalization of vision.

Conclusion: In a patient with a nonsecreting unilateral paraganglioma intracranial hypertension would be unusual; clinicians should remain vigilant for concomitant pathology in a symptomatic patient.

BACKGROUND

Glomus jugulare tumors are benign neoplasms that arise from the paraganglion cells in the adventitia of the jugular bulb and account for 60-80% of tumors of the jugular foramen.¹,² With a peak incidence in the 4th and 5th decades, paragangliomas are more common in females and may be sporadic or familial.¹ These tumors are highly-vascularized and often locally aggressive despite being histologically benign. Jugular foramen tumors most often present with pulsatile tinnitus and conductive hearing loss, but may also manifest aural fullness, dizziness, and lower cranial nerve palsies.¹,² Catecholamine secretion is extremely rare in head and neck paragangliomas but can lead to vasoactive symptoms such as palpitations, sweating, and headache.¹,²

Physical exam identifies a retrotympnic mass in 91% of glomus jugulare patients.³ High-resolution CT and MRI often give adequate information for diagnosis; therefore biopsy is rarely needed for confirmation.¹ Treatment options generally include observation, surgery, and either fractionated radiotherapy or stereotactic radiosurgery but should be individualized based on patient preference, symptoms, age, and comorbidities, as well as tumor size, extension, and growth rate.¹,²,³

In order to highlight a potential diagnostic conundrum, we present the case and subsequent management of unilateral paraganglioma with simultaneous contralateral sigmoid sinus stenosis causing bilateral outflow obstruction and leading to intracranial hypertension.

CASE PRESENTATION

A 58-year-old female presented with progressive right-sided pulsatile tinnitus, aural fullness, and mild conductive hearing loss. Physical exam demonstrated a red, pulsatile mass in the inferior quadrant of an intact right tympanic membrane. CT and MRI confirmed a right-sided glomus jugulare tumor measuring 2.0 x 1.9 cm within the jugular fossa causing right jugular bulb stenosis at the skull base and extending into the floor of the right middle ear (figure 1). Plasma-free and 24-hour urine-fractionated metanephrines were within normal limits, ruling out catecholamine secretion. CyberKnife stereotactic radiosurgery was completed for the right-sided glomus jugulare.

The patient began having progressive headaches and blurred vision with obscurations. Concern for intracranial hypertension, which would be unusual in the case of a slow onset blockage of a unilateral jugular bulb, led to additional testing including ophthalmologic examination that demonstrated early papilledema. Lumbar puncture confirmed intracranial hypertension with elevated opening pressure and transient alleviation of her headaches. Subsequent cerebral venography led to the discovery of a left-sided proximal sigmoid sinus stenosis with a pressure gradient of 25 mm Hg between the left transverse and sigmoid sinuses (figure 2). With validation of bilateral venous sinus stenosis causing intracranial hypertension, she was referred to interventional neuroradiology for consideration of dural sinus stenting.

Transfemoral catheterization with stenting across the left distal transverse and proximal sigmoid sinus led to immediate decrease in venous pressure gradients to 4 mm Hg with subsequent resolution of headaches and improvement in vision in the early postoperative period (figure 3). At 1 year follow-up, the patient reports continued symptom improvement and imaging demonstrates patent left dural venous sinuses with no progression of right-sided glomus jugulare.

DISCUSSION

Glomus jugulare tumors are the most common primary neoplasms at the jugular foramen and may cause venous outflow obstruction due to mass effect.¹ A presumptive clinical diagnosis can be made with a typical presentation of pulsatile tinnitus, conductive hearing loss, and a high-vascularity retrotympnic mass on exam. Cervical mass, other cranial nerve deficits, or rarely, vasomotor symptoms or headaches, sweating, palpitations, pallor, and nausea may also be indicative of a paraganglioma.⁴ The diagnosis of glomus jugulare is often confirmed on imaging with its contrast-enhancement with possible jugular foramen bony destruction on CT and T2-weighted hyperintensity with classic salt-and-pepper appearance on MRI.⁴ Depending on patient factors and tumor characteristics, treatment of these tumors can be straightforward and usually involves observation, surgery, and either fractionated radiotherapy or stereotactic radiosurgery.¹,³

Atypical symptoms in a patient with glomus jugulare, such as worsening headache and vision changes in this case, should alert the clinician to concomitant pathology and warrant additional workup. Catecholamine production by paraganglioma is essentially ruled out with normal plasma-free and 24-hour urine-fractionated metanephrines. Increased intracranial pressure was diagnosed on lumbar puncture but the etiology remained uncertain. Further imaging confirmed that bilateral dural venous sinus outflow was obstructed at the jugular bulb by mass effect and the sigmoid sinus by idiopathic stenosis. Once a critical point was met, proximal dural sinus pressures escalated and led to increased intracranial pressure and secondary headaches with visual disturbances.

Given our patient’s demographic, we also considered idiopathic intracranial hypertension (IIH) as a contributor to the additional symptoms. With an incidence of approximately 1 in 100,000, IIH is most common in obese women of childbearing age and can present with headaches, visual disturbances, aural fullness, hearing loss, and dizziness.⁵,⁶ Recent studies have found bilateral transverse sinus stenosis in greater than 90% of IIH patients although its controversial whether the stenosis is a result of and not the source of the elevated intracranial pressure.⁷ Additionally, one study has identified unilateral dural venous sinus stenosis in 30% of asymptomatic patients.⁸ Typical conservative measures for intracranial hypertension include acetazolamide, weight loss, and serial lumbar puncture, though recently, dural venous sinus stenting has become recognized as an efficacious treatment of intracranial hypertension in select patients with a pathologic pressure gradient associated with a stenotic segment.⁹,¹⁰

Our patient elected to forego conservative measures and undergo endovascular stenting of the proximal sigmoid sinus stenosis. This procedure was immediately successful in symptom improvement and avoided delay in definitive management of the increased intracranial pressure that resulted from bilateral venous sinus outflow obstruction. After stereotactic radiosurgery for the unilateral glomus jugulare and venous sinus stenting of the contralateral sigmoid sinus stenosis, the patient had continued symptom improvement, and no progression of tumor at 1 year follow-up.

CONCLUSIONS

• Atypical symptoms in a patient with glomus jugulare should alert the astute clinician to evaluate for concomitant pathology.

• Intracranial hypertension can result from bilateral sigmoid sinus outflow obstruction, but would be an abnormal sequela of a nonsecreting unilateral paraganglioma.

• Patients with a complex clinical presentation deserve a multidisciplinary approach.

• Immediate surgical intervention for the tumor should be postponed until other possible causes for increased ICP have been ruled out.

REFERENCES