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Large Glomus Tumor Presenting in a 9-Year-Old

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Keywords
- glomus
- paraganglioma
- children

Glomus tumor is a rare entity embryologically derived from neural crest cells. The most common location is the carotid body, followed by the jugular bulb. We present a sporadic case of a large skull base paraganglioma in a child presenting with unilateral otorrhea. University of Oklahoma Health Sciences Center Institutional Review Board exemption was obtained.

Case Report

A 9-year-old girl presented with 2 months of unilateral otorrhea. She was initially treated with antibiotic and steroid drops with no resolution. She subsequently developed granulomatous tissue that persisted despite conservative medical management and aural debridement. She had no pain, fevers, vertigo, or lower cranial nerve deficits. She had no previous history of childhood infections or ear surgeries. An audiogram showed a moderate to severe conductive hearing loss on the left. A computed tomography (CT) scan showed a large 3.5 × 3.5 × 3.7 cm expansile hypervascular mass within the jugular foramen causing erosion of the petrous bone and occiput with intracranial extension (Figure 1A). She was referred to our neurotologist for further evaluation. She was taken to the operating room, where a red, pulsatile mass pedicled to the anterior external canal was seen, with proclivity to bleed. Biopsy of the mass was performed, confirming paraganglioma. We obtained plasma catecholemines and an ultrasound of the kidneys, which were unremarkable. Magnetic resonance imaging (MRI) was performed showing a hypervascular mass expanding throughout the mastoid and protrusion into the posterior fossa. It was compressive inferiorly against the cerebellum and expanded downward, paralleling the jugular vein, to the C1-C2 level (Figure 1B).

The patient subsequently underwent a staged procedure involving a multidisciplinary team including a head and neck surgeon, neurotologist, and neurosurgeon. Day 1 included preoperative embolization. On day 2, a transmastoid approach with lateral temporal bone resection, transposition of the facial nerve, neck exploration with control of the major vessels, and retrosigmoid craniectomy was performed. Finally, a transotic approach with resection of glomus tumor was performed on the third day. The mass was resected in its entirety except for a small portion adherent to the carotid artery. Because of expectations of facial nerve weakness after mobilization/translocation, a gold weight implant was placed at the second-stage operation. She tolerated the procedures well, with only mild lower cranial nerve deficits, including mild tongue weakness. She had normal speech and swallow function evidenced by a normal dysphagogram on postoperative day 7. Her facial nerve function revealed an expected House-Brackman score of 6 after the second-stage surgery. Six weeks postoperatively,

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Figure 1. (A) Computed tomography scan with contrast. Left internal auditory canal, axial view. Note the expansile mass within the jugular foramen and compressing the cerebellum. (B) Magnetic resonance imaging, T1-weighted, axial view. Note the salt-and-pepper–appearing mass causing destruction of the left petrous temporal bone, filling the middle ear cavity, surrounding the otic capsule, and completely involving the jugular bulb.
she underwent proton-beam therapy, receiving a total of 45 Gy. She is currently doing well with good facial nerve tone.

Discussion
Glomus tumors are benign paragangliomas that arise from neural crest cells and occur most commonly in the head and neck. They usually present in the fourth to fifth decades as an asymptomatic, slow-growing neck mass. They can be sporadic or familial, isolated or multicentric, with malignant transformation rare. The familial types tend to develop at a younger age. A small percentage (1%-3%) of paragangliomas are functional, which release vasoactive substances into the systemic circulation. The patterns of growth include expansion within the temporal bone by means of preformed pathways that offer minimal resistance. For example, extension into the sigmoid sinus suggests posterior fossa involvement, as was seen in our reported patient. CT and MRI usually establish the diagnosis. The treatment of choice for most paragangliomas is surgical excision. However, if their location is in close approximation to important structures, there is increased risk of morbidity (cranial nerve X-XII deficits and vascular injuries) and mortality, which is estimated at 3% to 9%. Radiation therapy is an option for patients who are not surgical candidates.

In the literature, there is limited information on glomus tumors in children. Glomus tympanicum in infants has been reported, as well as childhood paragangliomas associated with familial inheritance. No other sporadic case of this size has been described in a child to our knowledge. Because of the rarity of these tumors in a child, we emphasize the importance of imaging studies after conservative measures fail, biopsy to establish diagnosis if feasible, and multistage/multispecialty involvement for adequate excision. The otolaryngologist should be aware of this entity as part of the differential diagnosis for refractory otorrhea.

Author Contributions
Kimberly K. Caperton, corresponding author, primary author; Wayne E. Berryhill, senior author.

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