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CASE REPORT

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Massive vagal schwannoma in an 11-year-old girl

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Abstract

We describe an unusual case of a young girl presenting with a large vagal schwannoma necessitating a transcervical-mandibulotomy approach for total tumor resection. The presentation is unique due to the size of the lesion, the patient's age, the operative approach, and molecular pathology.

KEYWORDS

head and neck surgery, neck masses, pediatrics, schwannoma, vagal schwannoma

1 | INTRODUCTION

Vagal schwannomas are rare peripheral nerve sheath tumors arising from the vagus nerve.[1] Typically diagnosed in adulthood at a median age of 44 years,[2] pediatric vagal schwannomas are exceedingly rare. Tumors may arise intracranially or extracranially, with the parapharyngeal space being the most common extracranial site in the head and neck.[3]

Surgical resection remains the treatment of choice for vagal schwannomas. Described operative techniques include gross total resection or intracapsular enucleation. However, the merits of either approach are dependent on the lesion size, location, and baseline nerve function.

We describe herein an unusual case of a young girl presenting with a large vagal schwannoma with an associated hypoglossal nerve palsy necessitating a combined transcervical-mandibulotomy approach for total tumor resection. The presentation is unique due to the size of the lesion, the patient's age, the operative approach required, and the paralysis of a cranial nerve that was not its nerve of origin.

2 | CASE REPORT

An 11-year-old girl presented with an asymptomatic, right neck mass first noted 7 months prior. Physical examination was notable for a large right parapharyngeal space mass on palpation of the oropharynx and neck, with medialization of the right oropharynx. Atrophy and fasciculations of the right side of the tongue were seen, consistent with a right hypoglossal nerve paralysis. Flexible fiberoptic laryngoscopy demonstrated a submucosal bulge in the right nasopharynx and oropharynx, with preserved bilateral vocal fold motion. All other cranial nerves were intact.

Magnetic resonance imaging (MRI) of the neck highlighted a large ovoid mass in the right parapharyngeal

space measuring 4.9 cm × 3.7 cm × 7.4 cm. The tumor extended from the jugular foramen superiorly to the level of C5 inferiorly, but without intracranial extension. The mass caused acute displacement of the internal and external carotid arteries anteriorly and compression of the internal jugular vein posterolaterally. T2-weighted MRI sequences demonstrated heterogeneous hyperintensity (Figure 1A,B). T1-weighted MRI sequences showed a hypointense lesion with homogeneous avid enhancement after administration of gadolinium-based contrast, consistent with schwannoma (Figure 1C).[1] Genetic testing and Pediatric Brain and Nervous System Tumor Panel (Invitae) were negative for any germline mutations. Following multidisciplinary review and extensive patient and family discussion, the decision was made to proceed with complete surgical excision, involving a combined transcervical-mandibulotomy approach (Figure 2A–E). Given this patient's young age, the large size of the lesion, its anatomically precarious location at the skull base, and her hypoglossal nerve paralysis, gross total resection was preferred to radiotherapy, enucleation, or observation to offer the best chance of long-term cure.

Histopathologic examination of the tumor confirmed schwannoma with immunohistochemistry showing diffuse S100 and focal SMA positivity. Memorial Sloan Kettering Integrated Mutation Profiling of Actionable Cancer Targets (MSK-IMPACT), a next-generation sequencing assay of all protein-coding exons of 468 cancer genes, demonstrated *TET1* deletion of exons 1 and 2.

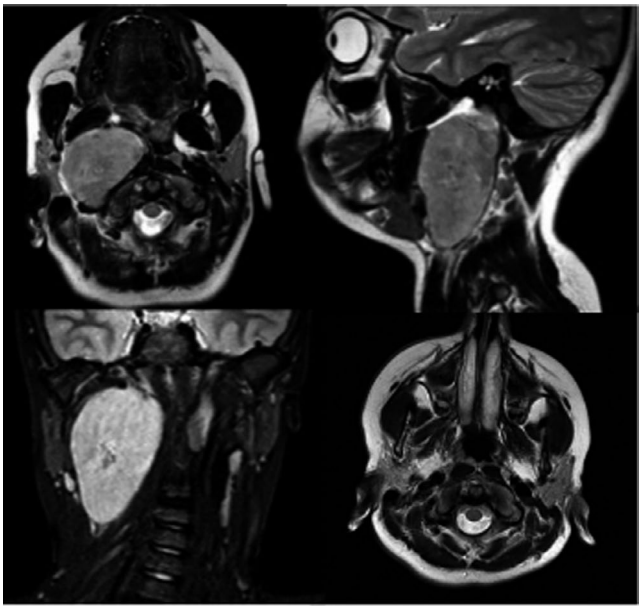

FIGURE 1 (A and B) T2-weighted MRI demonstrated a large ovoid mass in the right parapharyngeal space splaying the carotid artery and its branches and effacing the internal jugular vein. The tumor was heterogeneously hyperintense relative to skeletal muscle. (C) The lesion exhibits homogeneous avid enhancement after administration of gadolinium-based contrast, consistent with a radiologic diagnosis of schwannoma. (D) Imaging 6 months postoperatively demonstrates no evidence of disease

The patient underwent an uncomplicated postoperative recovery. The spinal accessory nerve was preserved. A temporary tracheostomy tube was removed prior to patient's discharge home. A temporary gastrostomy tube was assisted with nutritional supplementation during swallow rehabilitation. A right vocal fold injection augmentation with Prolaryn Plus (Merz Therapeutics) was performed and led to an improved voice. Postoperative coughing and throat clearing gradually improved. At 3 months after surgery, the patient was able to swallow without significant dysphagia or aspiration and a clear voice. MRI imaging at 6 and 12 months after surgery showed no evidence of persisting or recurrent schwannoma.

3 | DISCUSSION

Neurogenous tumors are rare among children, comprising only 2% of benign non-lymphadenomatous lesions.[4] Pediatric cervical vagal schwannomas are especially rare. The median age of diagnosis is 44 years in vagal schwannoma patients undergoing surgery.[2] The median maximum diameter of these tumors is 5 centimeters.[2] Cervical schwannomas may grow at varying rates ranging from 1 to 3 millimeters per year, and usually only become symptomatic once large enough to cause compressive symptoms.[1] Our patient's young age of 11 years, in tandem with the large size of the lesion, makes her presentation extraordinarily rare.

Schwannomas arise from a single nerve fascicle and may cause cranial nerve dysfunction by exerting pressure on other fibers of the affected or local cranial nerves. Reported series to date differ widely on the prevalence of cranial nerve deficits at presentation. Interestingly, our patient did not exhibit signs of vagus nerve dysfunction, despite the nerve being directly involved by the tumor, while she did have a long-standing hypoglossal nerve palsy, seemingly due to extrinsic compression from the vagal schwannoma. Although schwannomas do not typically present with cranial nerve palsies, when they do, dysfunction is usually of the index cranial nerve. Cranial nerve palsy may be suggestive of compression in a confined space or malignant schwannoma. Our patient's presentation with a hypoglossal nerve palsy with intact vagal nerve function in the context of a benign vagal schwannoma is very unusual.

Surgical resection is the preferred treatment modality for vagal schwannoma, either with gross total resection or with intracapsular enucleation. Gross total resection may be superior to enucleation with respect to cure, but often results in loss of ipsilateral vagal function distal to the

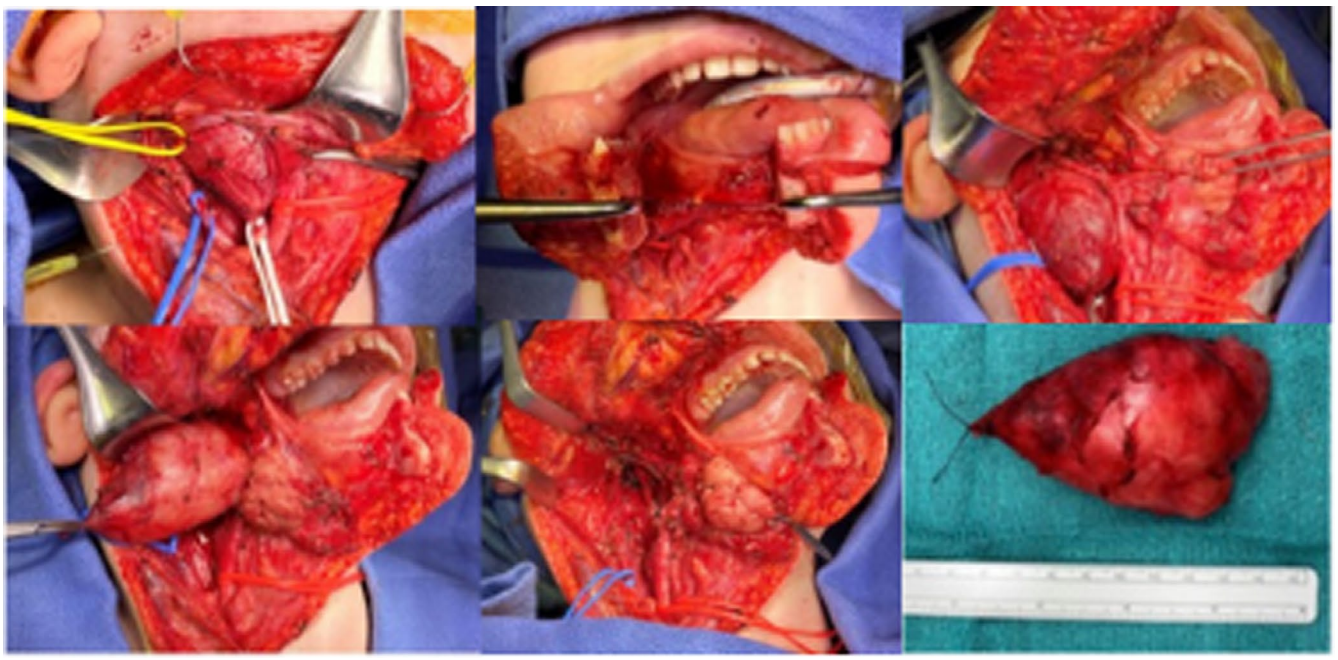

FIGURE 2 (A) The hypoglossal nerve (yellow vessel loop) is seen stretched and tented over the tumor, accounting for the patient's preoperative hypoglossal nerve paralysis. The vagus nerve (white vessel loop) can be seen coursing directly into the tumor. (B) A midline lower lip incision was used for a right paramedian mandibulotomy. The mandible was pre-plated before the osteotomy, and the dissection was then extended posteriorly inside the floor of mouth. (C) The jugular vein (blue vessel loop) is retracted anteriorly and the carotid artery (red vessel loop) courses anterior to the tumor. (D) Ligation and division of the external carotid artery with meticulous dissection over the tumor capsule at the skull base permitted a gradual delivery of the tumor. (E) Final operative site after tumor extirpation is shown. (F) Gross pathology specimen demonstrated a well-encapsulated, tan tumor 7.9 cm in greatest dimension

lesion. Enucleation may allow for preservation of nerve function, but is generally reserved for small schwannomas and is associated with higher rates of recurrence.

The transcervical approach is the most commonly employed technique to access vagal schwannomas.[2] Our combined transcervical-mandibulotomy approach was necessitated by an inability to safely gain access to the superior aspect of the tumor due to exension to the jugular foramen at the skull base and the adjacent, critical structures including the internal carotid artery, jugular vein, and spinal accessory nerve. Mandibulotomy allowed for improved access, safe mobilization of the tumor, and dissection of key structures at the skull base.

Vagal schwannoma can arise in the context of genetic syndromes, including neurofibromatosis type 2, Carney's complex, and schwannomatosis, with mutations in *NF2*, *SMARCB1*, *LZTR1*, and *PRKAR1a* previously described. Genomic changes in these genes all lead to downstream alterations in the production or function of the protein merlin, which plays a role in a range of growth factor pathways.[1] Our patient demonstrated no germline mutation or syndrome. Memorial Sloan Kettering Integrated Mutation Profiling of Actionable Cancer Targets (MSK-IMPACT) identified an isolated *TET1* deletion of exons 1 and 2. *TET1* plays a role in epigenetic tumorigenesis and may be implicated in schwannomatosis.[5] The clinicopathologic significance of the alteration in the setting of pediatric vagal schwannoma is unclear.

4 | CONCLUSION

We present a case of cervical vagal schwannoma that is rare in terms of the young age of the patient, the size of the lesion, the combined transcervical-mandibulotomy surgical approach taken, and the molecular pathology.

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CONFLICTS OF INTEREST

There exists no conflict of interest for any of the authors.

AUTHOR CONTRIBUTIONS

NZF, CWF, TM, and RJW provided the case report and approved of the final version to be published. They are accountable for the integrity of the content.

ETHICAL APPROVAL

I, Noah Feit, assure that the following are fulfilled: (1) This material represents original work not published elsewhere, (2) this paper is not currently considered for publication elsewhere, (3) the paper properly credits the contributions of all co-authors, (4) the patient history, surgical details, and outcomes are accurately described, (5) all sources are properly cited, and (6) all authors take responsibility for its content.

CONSENT

Informed written consent was obtained from the patient's parents for this publication.

MEETINGS

This manuscript has not been submitted to or presented at any meetings or conferences.

DATA AVAILABILITY STATEMENT

Data sharing not applicable – no new data generated: Data sharing is not applicable to this article as no new data were created or analyzed in this study.

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