Multimodal management of pediatric carotid body tumors: a systematic review and case illustrations

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OBJECTIVE Carotid body tumors (CBTs), extraadrenal paragangliomas, are extremely rare neoplasms in children that often require multimodal surgical treatment, including preoperative anesthesia workup, embolization, and resection. With only a few cases reported in the pediatric literature, treatment paradigms and surgical morbidity are loosely defined, especially when carotid artery infiltration is noted. Here, the authors report two cases of pediatric CBT and provide the results of a systematic review of the literature.

METHODS The study was divided into two sections. First, the authors conducted a retrospective review of our series of pediatric CBT patients and screened for patients with evidence of a CBT over the last 10 years (2007–2017) at a single tertiary referral pediatric hospital. Second, they conducted a systematic review, according to Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines, of all reported cases of pediatric CBTs to determine the characteristics (tumor size, vascularity, symptomatology), treatment paradigms, and complications.

RESULTS In the systematic review ($n = 21$ patients [includes 19 cases found in the literature and 2 from the authors' series]), the mean age at diagnosis was 11.8 years. The most common presenting symptoms were palpable neck mass (62%), cranial nerve palsies (33%), cough or dysphagia (14%), and neck pain (19%). Metastasis occurred only in 5% of patients, and 19% of cases were recurrent lesions. Only 10% of patients presented with elevated catecholamines and associated sympathetic involvement. Preoperative embolization was utilized in 24% of patients (external carotid artery in 4 and external carotid artery and vertebral artery in 1). Cranial nerve palsies (cranial nerve VII [$n = 1$], IX [$n = 1$], X [$n = 4$], XI [$n = 1$], and XII [$n = 3$]) were the most common cause of surgical morbidity (33% of cases). The patients in the authors' illustrative cases underwent preoperative embolization and balloon test occlusion followed by resection, and both patients suffered from transient Horner's syndrome after embolization.

CONCLUSIONS Surgical management of CBTs requires an extensive preoperative workup, anesthesia, and multimodal surgical management. Due to a potentially high rate of surgical morbidity and vascularity, balloon test occlusion with embolization may be necessary in select patients prior to resection. Careful thorough preoperative counseling is vital to preparing families for the intensive management of these children.

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KEYWORDS carotid body tumor; paraganglioma; embolization; oncology

CAROTID body tumors (CBTs), also known as paragangliomas, are rare vascular tumors arising from neuroendocrine cells within the carotid body. CBTs have a prevalence of 1:30,000 and represent more than 50% of head and neck paragangliomas.³¹ These tumors may produce significant symptomatology, including sympathetic surges from catecholamine release, palpable neck

mass, hoarseness, dysphagia, and Horner's syndrome.^{19,39} In the adult population, there is significant morbidity and mortality associated with the treatment of these lesions. While resection is the primary method of treatment for these tumors, this modality is technically challenging due to intimate associations with cranial nerves and the arterial system.⁹ Due to these surgical challenges, preopera-

ABBREVIATIONS BTO = balloon test occlusion; CBT = carotid body tumor; CN = cranial nerve; ECA = external carotid artery; ICA = internal carotid artery; MIBG = iodine-123-meta-iodobenzylguanidine; PRISMA = Preferred Reporting Items for Systematic Reviews and Meta-Analyses.

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tive embolization is widely utilized to provide surgeons with greater ease while reducing complications.³⁵ While CBTs most commonly affect adults in their 3rd to 5th decade of life, they are exceedingly rare in the pediatric population.³⁹ With relatively few pediatric cases reported in the literature (< 20 cases), surgical morbidity and optimal treatment paradigms for pediatric patients have yet to be established. Children may be especially vulnerable to these neoplasms due to their low intravascular volume, autonomic dysregulation, proximity to/infiltration of critical neurovascular structures, and potential associations with genetic syndromes.

Here, we present two new cases of pediatric CBT and present a systematic review of the literature to further understand the multimodal treatment of these lesions.

Methods

Systematic Review

Based on the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines, the authors performed a database search of PubMed to identify English-language literature published between 1968 and 2018, searching MeSH terms from PubMed, with combinations of keywords that describe pediatric paragangliomas, CBTs, and glomus tumor. Articles were limited to those written in English, those that included pediatric patients (under the age of 18 years), and those in which surgery was performed. Tumors were confirmed to be paragangliomas by histological classification based on features common to CBTs and adrenal paragangliomas. Articles were limited to those discussing tumors in the neck. Eighteen full-length articles were assessed; from these 4 were excluded and 14 were included in this analysis. Cross-referencing articles found by the initial search yielded no additional relevant articles (Fig. 1).

Results

Through our literature review, we identified 19 cases over the last 50 years, plus an additional 2 new cases from our institution, for a total of 21. The mean age at diagnosis was 11.8 years and there were a greater number of females. All patient demographics are listed in Table 1. A palpable neck mass was seen in 62% (n = 13) of patients, with tumors most commonly located in the right carotid body (62%). Of the 16 patients with tumor size reported, 44% presented with large tumors (> 5 cm in diameter); see Table 2. Most patients in our series had a successful gross-total resection (n = 18), one had subtotal resection, one received chemotherapy, and one underwent radiotherapy. Preoperative embolization was used in 24% of the cases; no cases of preoperative embolization resulted in any complications. The surgical morbidity rate was 33% (cranial nerve [CN] palsy, n = 7). Recurrence was noted in 19% (n = 4) of patients. Metastasis was evident only in one case (5%) and two patients displayed elevated catecholamine levels (10%). Overall, 66% of patients were documented to have follow-up with no evidence of disease or complications after the first treatment. Overall survival was 95.2%, with a mean follow-up duration of 39.5 months.

FIG. 1. PRISMA flow diagram. Figure is available in color online only.

Illustrative Cases From Our Institution

Case 1

This 8-year-old girl presented with a nontender palpable neck mass. She first noticed the mass after suffering recurrent streptococcal pharyngeal infections. The child reported no associated weight loss, dysphagia, hoarseness, fever, or signs of sympathetic surges. CT and MRI revealed a right cervical neck mass at carotid bifurcation without visualization of the right internal jugular on CT angiography. An MIBG scan (iodine-123-meta-iodobenzylguanidine nuclear scan) was negative for neuroendocrine tumor, and therefore the patient underwent a needle biopsy that yielded results consistent with paraganglioma. Prior to resection, she underwent diagnostic angiography with a balloon test occlusion (BTO) of the right internal carotid artery (ICA). The patient passed the BTO and had an uneventful embolization of her external carotid artery (ECA) feeders (Fig. 2). The patient was placed under general endotracheal anesthesia and the tumor was resected. Intraoperatively, we noted bleeding, and also noted was gross infiltration of the ICA lumen, which required parent vessel sacrifice. Collateral feeders from the ECA and vertebral artery were also ligated to facilitate resection. Intraoperatively, mild electroencephalogram slowing was noted, and somatosensory evoked potentials remained stable (estimated blood loss < 50 ml). Postoperatively, the patient developed mild hoarseness (CN X) and right-sided Horner's syndrome. Over the ensuing weeks, the transient vagal neuropathy and Horner's syndrome improved. Follow-up after 2 years showed resolution of the Horner's syndrome and vagal neuropathy and no further complications.

TABLE 1. Review of literature on pediatric CBTs

Authors & Year	Pt No.	Sex	Age (yrs)	Tumor Laterality	Tumor			Preop Elev Cat	Tx	Metastasis	Postop CN Palsy	Recurrence	Notes
					Size (cm)	Palpable Neck Mass	Embol						
Chambers & Mahoney, 1968	1	F	9	Rt	2	Yes*	—	—	GTR	—	—	—	
	2	M	12	Lt	2.5	Yes*	—	—	GTR	—	—	—	Heavy bleeding
	3	M	14	Rt	6	Yes	—	—	GTR	—	—	—	
Newland & Hurlbert, 1980	4	F	13	Lt	8	—	—	Yes	GTR	—	—	—	
Dickinson et al., 1986	5	F	12	Rt	—	—	—	—	GTR	—	Yes (CN XII)	—	
	6	F	17	Rt	—	—	—	—	GTR	—	Yes (CN XII)	—	
Thompson & Cohen, 1989	7	F	13	Rt	5	Yes	—	—	GTR	—	—	—	
	8	F	14	Bilat	2	—	—	—	GTR	—	Yes (CN VII)	—	Deafness in rt ear postop
Ophir, 1991	9	F	12	Bilat	4	—	—	—	GTR	—	Yes (CN IX, X, XI, & XII)	Yes	Op on recurrent tumor 6 mos; postop; NED after 8 yrs
Hajnić et al., 1999	10	M	9	Rt	5	Yes*	—	—	Chemo	Yes	—	—	Expired in 28 mos
Tekautz et al., 2003	11	M	8	Rt	2.5	Yes*	—	—	GTR	—	—	—	
	12	F	15	Rt	6.3	—	—	—	GTR	—	—	Yes	Subsequent Tx w/ radiotherapy; alive after 13 mos
Zaupa & Höllwarth, 2007	13	M	15	Rt	4	Yes	Yes (ECA)	—	GTR	—	—	—	
Georgiadis et al., 2008	14	F	13	Rt	0.5	Yes	—	—	GTR	—	—	—	
Fennessy et al., 2011	15	M	18	Bilat	2.1	—	Yes (ECA)	—	GTR	—	—	—	
Bensaid et al., 2015	16	M	3	Rt	8.5	Yes	—	—	Radio	—	—	Yes	55% shrinkage; pt well at 9 mos
Manjaly et al., 2015	17	F	3	Rt	—	Yes	—	—	STR (90%)	—	Yes (CN X)	Yes	Monitored using surveillance MRI w/o any immediate action
Ifoluwa et al., 2017	18	F	8	Lt	5	Yes	Yes (ECA)	—	GTR	—	—	—	Febrile for 3 days
Bakshi & Kumar, 2018	19	F	17	Lt	3	Yes	—	—	GTR	—	—	—	
Current series	20	F	8	Rt	—	Yes	Yes (ECA/VA)	—	GTR	—	Yes (CN X)	—	
	21	M	15	Lt	—	—	Yes (ECA)	Yes	GTR	—	Yes (CN X)	—	

Cat = catecholamines; Chemo = chemotherapy; Elev = elevated; Embol = embolization; GTR = gross-total resection; NED = no evidence of disease; Pt = patient; Radio = radiotherapy; STR = subtotal resection; Tx = treatment; VA = vertebral artery; — = not applicable.

* Indicates painful palpable neck mass.

TABLE 2. Baseline characteristics of 21 subjects

Pt Characteristics	Value
Age (IQR), yrs	11.8 (3–18)
Sex, no. (%)	
Male	13 (62%)
Female	8 (38%)
Presenting symptoms, no. (%)	
Painless neck mass	9 (43%)
Painful neck mass	4 (19%)
Cough/dysphagia	3 (15%)
Elevated catecholamines	1 (5%)
Tumor size, no.	
<5 cm in diameter	9
≥5 cm in diameter	7
Laterality of tumor, no. (%)	
Rt carotid body	13 (62%)
Lt carotid body	5 (24%)
Bilat	3 (15%)
CN palsy postop, no.	
VII	1
IX	1
X	4
XI	1
XII	3
Preop embol, no.	
ECA	4
ECA & VA	1

IQR = interquartile range.

Case 2

This 15-year-old boy presented for consultation for right-sided facial flushing with hyperhidrosis. He was noted to have left-sided Horner's syndrome as well as elevated urine metanephrines. An MIBG scan was negative for neuroendocrine tumor. After further evaluation, MRI/MR angiography revealed a left-sided neck mass along the left carotid artery displacing the carotid artery and the ECA and ICA anterolaterally. When the patient's Horner's syndrome and CN X neuropathy worsened, it was determined that the patient would benefit from preoperative embolization followed by resection of the mass. Preoperatively, the patient underwent embolization and BTO through the superior ECA feeders. The following day, he was taken to surgery, placed under general endotracheal anesthesia, and the operation proceeded with no complications (estimated blood loss < 50 ml). Tight blood pressure control was advised during this procedure. The common facial vein, large feeders coming from the inferior pole of the tumor, and one superior feeding vessel were divided during the operation with use of a microscope for visualization. The entire tumor was removed, and histopathology confirmed the diagnosis of paraganglioma (Fig. 3). The patient is doing well at 2 years following treatment with an improving left-sided Horner's syndrome.

Discussion

CBTs are rare tumors that account for a very small portion of all neoplasms in the head and neck region. The tumor was first described by Albrecht von Haller in 1743.²⁴ CBTs are normally slow-growing and are generally asymptomatic for long periods of time. The majority of patients present with unilateral tumors, but 3%–4% can be bilateral.²² Due to their slow growth, CBTs may go unnoticed for long periods of time. Symptoms are eventually caused by the mass of the lesion or by the lesion's involvement of cranial nerves. Common symptoms include neck pain, hoarseness, dysphagia, Horner's syndrome, cough, and tongue weakness.^{1,3} The tumor mass exhibits only medial-to-lateral mobility and unique pulsations can be felt immediately anterior to the lesion.^{3,25} Cranial nerve involvement is quite rare, only occurring up to 20% of the time, often involving the glossopharyngeal, hypoglossal, and vagal nerves.^{16,41}

Etiology

The specific cause of these tumors is unknown. However, a higher prevalence of CBTs has been identified in people residing at higher altitudes in hypoxic conditions. Saldana et al. reported that Peruvian patients residing at high altitude presented with carotid body hyperplasia 10 times more frequently than individuals at sea level.³⁶ Low pO₂ is a major determinant of chemoreceptor cell activity, and thus it has been postulated that hypoxic conditions may be a stimulus for carotid body hyperplasia. In addition, several genes have been suspected in the development of these tumors, such as *NFI*, *RET*, *SDHB*, and *TMEM127*.²⁸ It has been suggested that individuals who carry CBT susceptibility mutations and are subjected to chronic hypoxia often develop tumors at an earlier age.²⁰

Diagnosis

The differential diagnosis for these lesions commonly includes enlarged lymph nodes, branchial cleft cysts, parotid gland tumors, thyroid tumors, neurogenic tumors, and carotid artery aneurysms.³ The combination of classic clinical symptomatology (hoarseness with sympathetic dysregulation) and radiological findings (tumor in the carotid bifurcation) can help seal the preoperative diagnosis.⁷ Newer techniques, such as MR angiography, have also been utilized for accurate diagnosis and preoperative planning.¹⁴ Ancillary radiological studies, such as positron emission tomography (PET) scanning or nuclear medicine MIBG, may also be helpful in identifying foci of increased metabolism or neuroendocrine production, respectively.¹⁴ Additional tests such as urine metanephrine testing should also be performed if a diagnosis of CBT is suspected, although these tests can show normal findings, as indicated in our case illustrations. Given their high vascularity and proximity to large neurovascular structures, incisional biopsies are usually not recommended.¹³ In case 1, a needle biopsy was performed due to the uncertainty of diagnosis in the setting of negative adjuvant tests (negative MIBG and urine metanephrines).

Anesthesia

Preoperative management of blood pressure is impor-

FIG. 2. An 8-year-old girl with a neck mass. Sagittal (A) and coronal (B) MR images demonstrating a 3.4-cm contrast-enhancing lesion at the carotid bifurcation. Lateral (C) and anterior (D) views of right common carotid artery cerebral angiograms demonstrating arterial blush within lesion with ECA and ICA feeders. BTO of the right ICA (E) demonstrating adequate filling of right ICA. Postoperative coronal MR image (F) demonstrating gross-total resection of cervical lesion.

tant for limiting surgical complications and should be considered in all operative cases. Although in most cases the tumors are not actively secreting, some paragangliomas secrete catecholamines, making the risk of surgery greater. Whether or not the tumor is actively secreting catecholamines, there is a risk that catecholamine surge can occur during resection and lead to severe vasoconstriction through alpha receptors and cause a hypertensive crisis. Thus, in order to prevent this from occurring, it is important to utilize alpha blockers preoperatively, such as phenoxybenzamine. Phenoxybenzamine is a robust blocker that is able to inhibit alpha activation and is not easily overcome by any catecholamine surge.¹¹ Following adequate alpha blockade, beta blockers may also be utilized to control heart rate.¹² Adequate blood pressure control is important to avoid hypertensive crises during surgery.

Treatment

Typical treatment of these lesions requires surgical resection coupled with preoperative angiography and potential embolization. For children, dedicated angiography may help delineate important vascular feeders, and embolization may prevent excessive blood loss during surgery.^{32,42} After embolization of vascular feeders, reduced intraoperative blood loss may improve the surgeon's visibility and reduce the risk of inadvertent nerve injury.^{21,34,43} During angiography, a BTO can be performed, which can determine whether the patient can tolerate a carotid artery sacrifice during surgery or preoperatively. It is critically important to evaluate ischemic tolerance using BTO to prevent stroke. BTO results are most commonly evaluated during either clinical or radiological examination. In the clinical evaluation method, hand grip, muscle strength,

language, memory, and facial strength are assessed several times over a 30-minute period. On the other hand, adjuvant imaging can be used to measure the symmetry of arterial/venous filling and determine whether there is sufficient collateral blood flow.³⁸ These evaluations are important when designing the patient's treatment plan to include embolization. Although not always necessary, embolization may provide an additional safeguard for resection of these difficult tumors in children. However, the added benefit of preoperative embolization must be weighed against its po-

FIG. 3. Immunohistochemistry on formalin-fixed, paraffin-embedded tissue was used for diagnosis. Type 1 chief cells stain positively for synaptophysin, which confirms the neuroendocrine origin and supports the diagnosis of paraganglioma. Figure is available in color online only.

FIG. 4. Treatment paradigm for CBT management. CTA = CT angiography; MRA = MR angiography; VA = vertebral artery.

tential risks. In the setting of a failed BTO, the aggressiveness of embolization and the subsequent resection should be tailored accordingly. As in case 1, Horner's syndrome may develop after embolization; however, more serious complications have been reported with particles, including embolic stroke and postembolization inflammatory swelling.^{29,45} In children, clinicians should be wary of these risks as they counsel patients prior to surgery.

Gross-total resection is often considered the mainstay optimal treatment choice for CBTs. It has been reported

that recurrence is highly unlikely when gross-total resection is achieved.²⁷ In our series, recurrence was only identified in 5% of pediatric cases who underwent gross-total resection with 6 weeks of follow-up. During surgery, early identification of critical neurovascular structures (ECA, ICA, jugular vein, and vagus nerve) is essential prior to debulking. Since these tumors may be intimately involved in the adventitial layers of the carotid artery, caution must be given to defining a plane or "white line" between the tumor and the artery.^{5,18} Once a diagnosis is suspected, early

and aggressive resection is recommended to prevent tumor growth, malignant degeneration, and avoid morbidity.³³

The ability for total resection is dependent on the size of the tumor and degree of attachment to the carotid vessel. Shamblin classification (types 1–3) has helped guide resection: type 1 (relatively small, minimally attached), type 2 (larger, moderately attached), and type 3 (large with carotid incarceration). Type 3 CBTs are difficult to resect and may require vascular sacrifice in order to obtain a gross-total resection.³⁷

Limitations

This study is inherently limited due to publication and selection bias of any review of the literature. These studies may underreport complications and may not accurately represent surgical morbidity of pediatric paragangliomas. Lastly, the mean overall follow-up in our systematic review was 39.5 months; therefore, a true estimate of recurrence risk is difficult to predict.

Conclusions

CBTs in a pediatric population can cause significant morbidity and mortality if untreated, but the tumors usually have a good prognosis with surgical interventions. Most tumors present as asymptomatic palpable neck masses but can cause morbidity due to mass effect or secretion of catecholamines that cause increased blood pressure, palpitations, and cardiac arrhythmias. This review shows that metastasis in this tumor type is rare and that surgical intervention has been successful as the primary treatment method. Our case series demonstrates that preoperative embolization is beneficial in the management of CBTs by reducing intraoperative bleeding, which may lead to better resection and fewer complications. Our treatment paradigm can be seen in Fig. 4.

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Disclosures

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Conception and design: Shah, Jernigan. Acquisition of data: Niazi. Drafting the article: Kuchakulla, Shah, Armstrong. Critically revising the article: Bhatia.

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