Pediatric carotid body tumors: A case report and systematic review

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ABSTRACT

Carotid body tumors (CBTs), or chemodectomas, are rare, especially in the pediatric population. They often present with minimal symptoms, making timely diagnosis challenging. This case report and systematic review highlights a distinctive presentation and summarize the current evidence on pediatric CBTs. We report a case of a 13-year-old girl presenting with neck pain and a left-sided neck mass. After extensive evaluation, a Shamblin type III tumor was identified and removed surgically. Postoperatively, the patient experienced transient hypertension and significant dysphagia, both of which resolved within a few weeks with no permanent sequelae. Histology confirmed a benign paraganglioma. A systematic literature review of PubMed identified 29 cases from 23 published studies spanning from 1968 to 2024. The average age at diagnosis was 12.6 ± 3.6 years. The most common symptom was a neck mass or swelling, reported in 75% of cases (n = 21). Tumor sizes ranged from 1.3 to 8.0 cm, with Shamblin III being the most frequent classification. Gross total resection (n = 25 [89.3%]) alone or in combination with preoperative embolization (n = 10 [35.7%]) were the most common methods of management. In 62.1% of cases, there were no permanent complication or sequelae. The proximity to vital neurovascular structures and high vascularity in pediatric patients necessitates careful perioperative interdisciplinary management. Owing to their rarity and nonspecific presentation, CBTs often remain undiagnosed for years. They respond well to treatment, but can be fatal if untreated, underscoring the importance of including CBTs in the differential diagnosis of pediatric neck masses. (J Vasc Surg Cases Innov Tech 2024;10:101584.)

Keywords: Carotid body tumor; Paragangliomas; Chemodectomas; Neck tumor; Balloon test occlusion

Paragangliomas are rare neuroendocrine tumors originating from the extra-adrenal chromaffin cells of the autonomic nervous system and are most commonly found in the head, neck, or abdomen.[1,2] Owing to their unique features, clinical presentation varies based on location and hormone-secretory activity.[3] Carotid body tumors (CBTs) are a subset of head and neck paragangliomas located within the adventitia of the carotid body at the bifurcation of the carotid artery.[1] CBTs are highly vascular, accounting for 0.3% of all paragangliomas and 60% of head and neck paragangliomas.[4] Also known as chemodectomas, they primarily consist of neural crest-derived cells and are more common in middle-aged women.[5] These slow-growing lesions are typically fixed vertically to the carotid bifurcation—a phenomenon known as Fontaine's sign.[4] Most CBTs are asymptomatic, but they can cause disturbances such as pain, tongue paresis, hoarseness, Horner's syndrome, and dysphagia owing to mass effect.[4] Hormone-secreting CBTs can present with signs and symptoms such as paroxysmal hypertension, palpitations, headaches, dizziness, flushing, diaphoresis, or photophobia.[5] Approximately 6% of CBTs are malignant, with the potential for lymphatic spread or distant metastases.[6] CBTs are extremely rare in children and adolescents; the last systematic review, published in 2018, reported 21 patients in this age group,[7] with additional cases reported since then. Given this background, we report a case of CBT in a 13-year-old patient and present the findings of an up-to-date systematic review.

CASE REPORT

A 13-year-old girl sought our attention for a left-sided neck mass persisting for 2 years. Initially asymptomatic, she began experiencing intermittent mild-to-moderate neck pain (scoring 3-4 on a 0-10 numeric rating scale) a few months before her presentation. Her medical history included neonatal hydrocephalus managed with a ventriculoperitoneal shunt and bronchial asthma controlled with albuterol. She resided in a city located at sea level and did not report a family history of CBT or paraganglioma. Despite seeking medical attention at various hospitals, the cause of her neck mass remained undiagnosed. A previous

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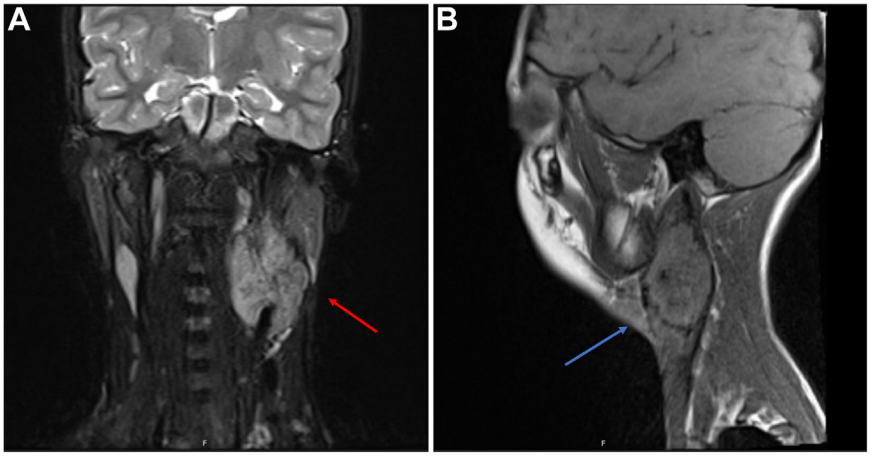

Fig 1. Coronal view **(A)** of magnetic resonance imaging and sagittal view **(B)** highlighting a Shamblin III carotid body tumor (CBT), measuring 4.3 × 3.6 cm, located at the bifurcation of the left common carotid artery (red and blue arrows).

needle biopsy of the cervical lesion at another hospital had revealed follicular hyperplasia and ruled out infectious causes.

Physical examination revealed a significant neck swelling, measuring 4.0 cm × 3.0 cm, without significant overlying skin changes. Ultrasound examination showed a 4.3 cm × 3.6-cm mass at the left carotid bifurcation. Computed tomography angiography suggested features indicative of a CBT, and subsequent magnetic resonance imaging classified the mass as Shamblin type III, indicating that it encompassed vascular structures (Fig 1, *A* and *B*). The distance to the base of the skull was 1.6 cm, and the tumor volume was 34 mL. Laboratory results were within normal ranges, and urinary metanephrine testing was negative. Preoperatively, the patient underwent a left carotid balloon occlusion test to assess brain perfusion in preparation for potential perioperative complete excision of the left internal carotid artery (ICA). The test was passed without displaying any symptoms (Fig 2, *A* and *B*).

Intraoperative neuromonitoring was used to monitor for possible neurological alterations. After exposure, the mass exhibited unusually high vascularity with friable vessels prone to bleeding. The common, external, and internal carotid arteries were identified clearly, and the external carotid artery was clamped to minimize bleeding, because it was the primary arterial feeder for the tumor. Extremely careful dissection of the ICA and carotid bifurcation was performed using bipolar electrocautery owing to the tumor's high adherence to blood vessels and adjacent cranial nerves. A complete en bloc surgical resection of the carotid body and ICA was achieved without macroscopic residual remnants (Fig 3). All nerve structures were visualized and preserved throughout the whole procedure. Owing to the small size and high location of the ICA near the base of the skull, combined with the unremarkable preoperative balloon occlusion test, we opted against performing vascular reconstruction.

Two units of packed red blood cells were transfused intraoperatively, and phenylephrine was administered to prevent hypotension from surgical blood loss. Postoperatively, the patient was monitored in the postoperative care unit for vital and neurological signs. She experienced hypertension, which was managed with clonidine and amlodipine.

Physical examination revealed a postoperative left tongue deviation owing to probable hypoglossal nerve palsy, and the swallowing team attributed it to pharyngeal dysphagia, recommending nasojejunal (NJ) tube placement. Histopathological analysis confirmed the diagnosis of a benign paraganglioma.

After receiving education on feeding, the patient was discharged home with an in situ NJ tube and an amlodipine prescription for hypertension. At the 4- and 6-month follow-ups, neurological symptoms had improved significantly, the NJ tube was removed, and she returned to her baseline, with an unremarkable recovery. Owing to gradual improvement of her blood pressure, amlodipine was discontinued gradually from 6 months postoperatively. Genetic testing was offered to the patient's family; however, they chose not to pursue further evaluation.

The child's parent provided written informed consent for the publication of their daughter's case.

METHODS

Search and selection. We conducted a systematic review of all published pediatric CBT cases. The search and reporting were performed in compliance with the PRISMA guidelines.[8] The PubMed database was searched from inception until January 2024 using a keyword combination to capture all CBT cases using a mixture of medical subject headings, free text words, and word variants combined with appropriate Boolean operators: (("carotid body tumour" [All Fields] OR "carotid body tumor" [MeSH Terms] OR ("carotid" [All Fields] AND "body" [All Fields] AND "tumor" [All Fields]) OR "carotid body tumor" [All Fields]) OR ((("neck"[MeSH Terms] OR "neck"[All Fields]) AND ("paraganglioma"[MeSH Terms] OR "paraganglioma"[All Fields] OR "paragangliomas"[All Fields])). To supplement the electronic search with a

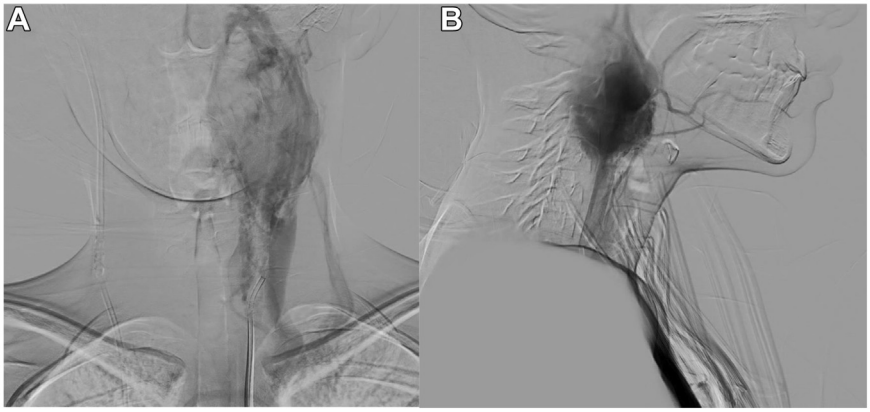

Fig 2. Coronal view **(A)** of preoperative balloon occlusion test and sagittal view **(B)** displaying a highly vascularized tumor located at the bifurcation of the left common carotid artery.

Fig 3. Surgical specimen of the resected carotid body tumor (CBT) measuring approximately 5 × 3 cm.

snowballing method, we screened the bibliographies in the published literature. This search was limited to case reports. The inclusion criteria for the synthesis were the pediatric age group and case reports published in the English language. Cases that included patients >18 year old were excluded. After screening the title and abstract, articles were obtained for full-text assessment. Studies with inaccessible full texts were excluded at this stage. Publications that met all the eligibility criteria were used for data synthesis. PRISMA flowchart was generated using a Shiny app by Haddaway et al.[9]

Data synthesis. Of the 1688 records screened initially, 29 full texts were available for screening. Of these, six were excluded for the following reasons: missing outcome of interest in pediatric cases (n = 4)[10-13] and publication language other than English.[14,15] The selected studies included a total of 29 cases from 23 studies reported between 1968 and 2024[7,16-37] (Table). The search and selection processes are presented in PRISMA flow chart (Fig 4).

A large number of cases were reported from the United States (n = 13 [44.8%]), followed by Greece (n = 3 [10.3%]) and India (n = 3 [10.3%]). The average age of children at diagnosis was 12.6 ± 3.6 years (range, 3-18 years). The majority of patients were female (n = 18 [62.1%]) and, among the reported cases, they mainly were of Caucasian race. Two cases had a family history of paraganglioma,[21,26] and one patient had an *SDH* gene mutation.[36] Furthermore, a history of B-cell acute lymphoblastic leukemia,[22] type 1 diabetes mellitus,[24] and congenital agenesis of the external ear was reported in individual cases, whereas gastric leiomyosarcoma and intrapulmonary paraganglioma[18] were reported in another case subsequently. Two patients lived at an altitude of 1500 m above sea level, and one of them experienced increased growth of the lesion after moving to a higher altitude region.[25,27]

The most common symptom that led to a diagnosis was neck mass or swelling (n = 21 [75%]). Neck masses were mostly nontender, and only four patients (13.8%) reported a painful swelling.[16,23,35] In several cases, neck swelling was misdiagnosed as cervical lymphadenopathy or brachial cleft cyst and was observed from a few months up to 8 years.[25,33] In two cases, a mass was detected in the ear canal during a routine examination, and in one of them a diagnostic biopsy resulted in uncontrollable bleeding.[20,26] Neurological deficits and compressive symptoms, such as hearing loss, deficits in the III to VI and X cranial nerves, and Horner's syndrome, were the next commonly observed findings that led to a diagnosis. The lesion size varied from 1.3 to 8.0 cm. Among the reported cases, Shamblin III was the most common tumor type.[25,27,28,30,35] In almost one-half of the patients, the tumor was located on the right side

Table. Characteristics of the included studies and patients

Author, year	Publication year	Country	Age	Sex	Race	Symptoms	Size, cm	Side
Chambers et al.[16] (Patient 1)	1968	USA	12	Male	White	Mass, pain, syncope	2.5 × 2.5	Left
Chambers et al.[16] (Patient 2)	1968	USA	14	Male	Black	Mass, dysphagia	6 × 6	Right
Chambers et al.[16] (Patient 3)	1968	USA	9	Female	White	Mass, pain	1.5 × 2.0	Right
Newland et al.[17]	1980	USA	13	Male	White	Heart murmur, multiple joint pain and swelling, low-grade fever, weight loss	2.5 × 8.0	Left
Carney et al.[18]	1983	USA	12	Female	White	Asymptomatic mass	N/A	Left
Dickinson et al.[19] (Patient 1)	1986	UK	17	Female	–	N/A	N/A	Right
Dickinson et al.[19] (Patient 2)	1986	UK	12	Female	–	N/A	N/A	Right
Thompson et al.[20]	1989	USA	14	Female	White	Mass in the right ear canal detected in a routine examination	2 × 2	Bilateral
Ophir et al.[21]	1991	Israel	12	Female	–	Hearing loss, pulsatile tinnitus in the right ear for 6 months	N/A	Bilateral
Tekautz et al.[22] (Patient 1)	2003	USA	8.2	Male	White	N/A	2.5 (greatest dimension)	Right
Tekautz et al.[22] (Patient 2)	2003	USA	15.5	Female	White	Deficit in the right III to VI cranial nerves	6.3 (greatest dimension)	Right
Osborne et al.[23]	2005	USA	16	Female	–	Painful, nonpulsatile neck mass Shooting head pain upon fine-needle aspiration biopsy	3	Left
Zaupa et al.[24]	2007	Austria	15	Male	White	Asymptomatic immobile mass in the right side of the neck	4.0 × 3.0 × 2.5	Right
Georgiadis et al.[25]	2008	Greece	13	Female	–	Painless, pulsatile, slowly growing mass since the age of 9 years	5.0 × 4.3 × 3.0 (Shamblin III)	Right
Fennessy et al.[26]	2011	USA	18	Male	–	An asymptomatic red mass in the right external auditory canal was discovered at a routine examination	Left: 2.5 × 1.8 × 2.8; Right: 1.3 × 0.9	Bilateral
Lopez-Vazquez[27]	2014	Mexico	16	Female	–	Asymptomatic growing mass on the left side of the neck for the last 1 year	4 × 3 × 3 (Shamblin IIIb)	Left
Bensaid et al.[28]	2015	Morocco	3	Male	Middle East/ North African	Mass on the right side of the neck	8.0 × 5.3 × 4.5 (Shamblin III)	Right
Ifeoluwa et al.[29]	2017	Hungary	8	Female	–	Pulsating, progressively growing neck swelling for the last 2 years	5.0 × 4.1 × 5.8	Left
Singh et al.[30]	2017	India	13	Male	South Asian	Painless slowly (for 4 months) growing neck swelling on the left side	Left: 3.0 × 2.0 (Shamblin III); Right: Shamblin I	Bilateral
Bakshi et al.[31]	2018	India	17	Female	–	Progressively increasing swelling in the neck for the last 9 months	2 × 3	Left
Hogan et al.[32]	2018	USA	8	Female	–	Neck mass for the last 1 year	4.2 × 3.4 × 2.1	Right
Kotsis et al.[33] (Patient 1)	2019	Greece	16	Female	–	Mass on the left side of the neck	2.6 × 2.1 × 30 (Shamblin II)	Left
Kotsis et al.[33] (Patient 2)	2019	Greece	15	Male	–	Swelling on the right side of the neck (misdiagnosed as a brachial cleft cyst and followed up for 8 years)	6.0 × 3.5 (Shamblin II)	Right
Kuchakulla et al.[7] (Patient 1)	2019	USA	8	Female	–	Nontender palpable neck mass	N/A	Right
Kuchakulla et al.[7] (Patient 2)	2019	USA	15	Male	–	Left-sided Horner's syndrome, cranial nerve X neuropathy	N/A	Left
Formanek et al.[34]	2020	Czech Republic	13	Female	–	Painless mass in the right upper neck that had been growing for the last 7 years	4.0 × 3.0 × 3.5	Right
Abderrahim et al.[35]	2020	Tunisia	15	Female	–	A slowly growing painful mass on the left side of the neck that developed over 1 year	3.5 × 4.0 cm (Shamblin III)	Left
Molina Vázquez et al.[36]	2021	Spain	8	Female	Maghreb origin	Painless mass for the last 4 months	3.0 × 2.5 (Shamblin I)	Right
Yadav et al.[37]	2024	India	9	Male	–	–	–	–

CBT, Carotid body tumor; *EBRT,* external beam radiation therapy; *GTR,* gross total resection; *N/A,* not applicable.

Table. Continued.

Management	Pathology	Follow-up notes	Other notes
GTR	Benign CBT	No evidence of disease after 10 years	
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GTR	Carotid body paraganglioma (chemodectoma)	Postoperative chemotherapy and radiotherapy owing to incomplete resection	
N/A	CBT	Congenital agenesis of left and partial agenesis of right external ear. Diagnosis of gastric leiomyosarcoma and intrapulmonary chondromas and paraganglioma at the age of 24 years	No relevant family history
GTR	CBT (unspecified)	Uneventful recovery	–
GTR and external carotid artery ligation	CBT (unspecified)	Permanent XII cranial nerve damage	–
GTR (right-side surgery was followed with the left-side surgery after 2 months)	Carotid tumor and a right glomus jugulare tumor	Temporary complete right facial paralysis with slight vertigo. Total deafness in the right ear. Pulsatile tinnitus in the left ear	–
GTR (right-side surgery was followed with the left-side surgery after 6 months)	Bilateral carotid body paraganglioma and right jugular paraganglioma	Postoperative paralysis of the IX to XII cranial nerves without hemiplegia Paralyzed vocal cords were recovered following rehabilitation Eight years follow-up without recurrence	Family history of cerebellar astrocytoma in father and mediastinal paraganglioma in mother
GTR	Carotid body paraganglioma	No evidence of disease after 9.8 years of follow-up	Pre-B cell acute lymphoblastic leukemia 11 months before the diagnosis
GTR	Carotid body paraganglioma (bone, central nervous system, right parotid gland invasion, and cervical lymph node involvement)	Progressive disease Local tumor progression within 6 weeks of tumor embolization and an EBRT treatment Alive with a disease after 13 months	Family history of breast cancer and brain tumor
Preoperative embolization + GTR	CBT	N/A	–
Preoperative embolization + GTR	Carotid body paraganglioma	No evidence of disease after 18 months of follow-up.	History of type 1 diabetes since the age of 8 years Altitude 400 m
GTR	Carotid body paraganglioma	No evidence of disease after nine months of follow-up.	Accelerated growth after relocation to an altitude about 1500 m above sea level No evidence of family association
Preoperative embolization + GTR (right-side surgery was followed with the left-side surgery after 9 months)	Bilateral carotid body paraganglioma and right glomus jugulare tumor (paraganglioma)	No evidence of disease after 1 years of follow-up	History of carotid body paraganglioma in brother (at 25 years of age) and father
GTR with dissection of vagal and glossopharyngeal nerves	Carotid body paraganglioma	Postoperative left palpebral ptosis, anisocoria with left myosis, and dysphonia	Mean altitude 1,570 m above sea level No relevant family history
Radiotherapy without surgery (owing to the locally advanced disease)	Carotid body paraganglioma	Shrinkage to 55% after radiotherapy (9 months of follow-up)	No relevant family history
Preoperative embolization + GTR	Benign CBT	No evidence of disease after 6 months of follow-up	No relevant family history
Left: Preoperative embolization + GTR Right: Observation	Carotid body paraganglioma (left)	No postoperative sequelae	No relevant family history No history of residing in hilly areas
GTR	CBT	No evidence of disease after 14 months of follow-up	–
Preoperative embolization + GTR	Carotid body paraganglioma	No evidence of disease after 6 months of follow-up	No relevant family history
GTR	Carotid body paraganglioma	Postoperative transient episode of left parietal hypoesthesia	No relevant family history
GTR	Carotid body paraganglioma	–	No relevant family history
GTR with embolization	Carotid body paraganglioma	No permanent postoperative sequelae	–
GTR with embolization	Carotid body paraganglioma	No evidence of disease after 1 years of follow-up Improvement in left-sided Horner's syndrome	–
GTR with embolization	Carotid body paraganglioma	No evidence of disease after 2 years of follow-up	No genetic basis was found
Lost to follow-up	Carotid body paraganglioma	Sudden death after one year from the loss from follow-up (cause of death: thoracic aortic dissection and pericardial tamponade, most likely owing to the secreting jugulocarotidian paraganglioma)	No relevant family history
GTR with embolization	Carotid body paraganglioma	N/A	*SDH* mutation Aunt with a history of unspecified cervical surgery
177 Lu-DOTATATE radionuclide therapy	Metastatic carotid body paraganglioma (cervical lymph nodes and lungs)	Complete response after four cycles of 177Lu-DOTATATE radionuclide therapy	–

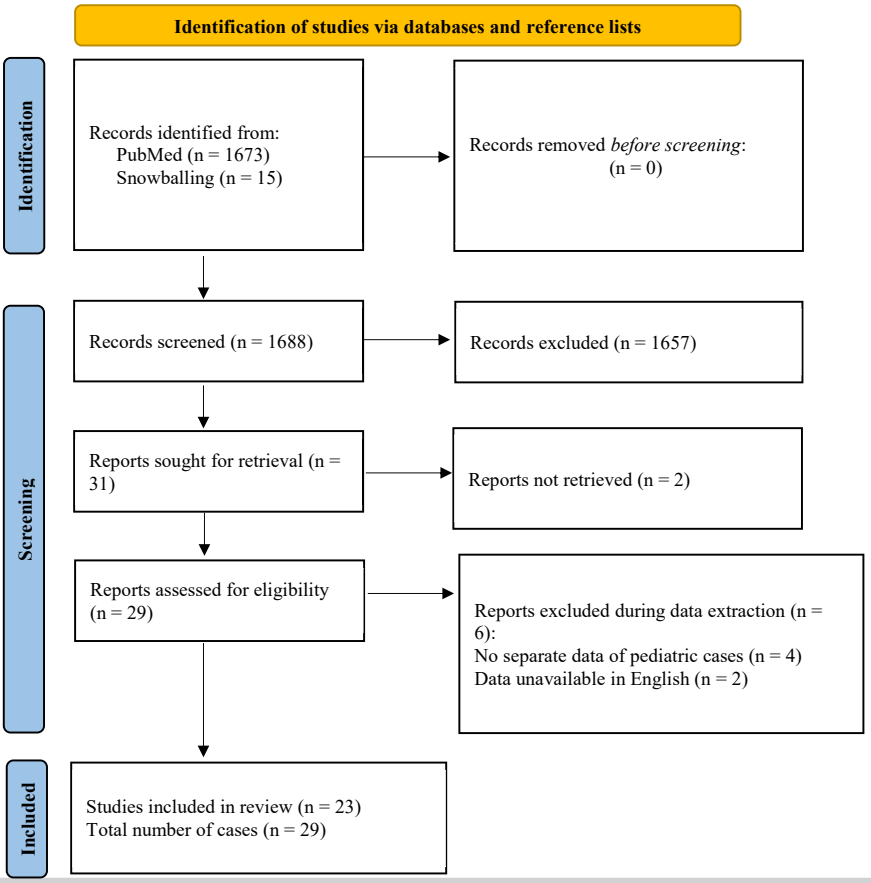

Fig 4. PRISMA flow diagram showing an overview of the study selection process.

(n = 14 [48.3%]), whereas, in four cases (13.8%), bilateral involvement was detected.[20,21,26,30] Interestingly, 75% of bilateral cases also had a right glomus jugular tumor,[20,21,26] and the remaining case presented with multiorgan and lymphatic involvement.[22]

Most cases underwent gross total resection (n = 25 [89.3%]), and 35.7% of cases (n = 10) received preoperative embolization. A patient with a Shamblin IIIb tumor required dissection of the glossopharyngeal and vagal nerves,[27] whereas another patient with incomplete resection underwent postoperative chemoradiotherapy.[17] Cases with bilateral tumors were operated in different sessions, with 2- to 9-month intervals. In one case, owing to the small size and Shamblin I type of the contralateral lesion, the tumor was left untreated.[30] Owing to the locally advanced nature of the disease, one case was considered inoperable and treated with radiotherapy,[28] and another patient with lung metastases and cervical lymph node involvement was treated with 177 Lu-DOTATATE radionuclide therapy.[37] Furthermore, one patient was lost to follow-up after a diagnostic workup.[35]

In the majority of patients, postoperative histology reports confirmed carotid body paraganglioma or CBT, and the benign nature of the tumor was defined in only four patients.[16,29] In the postoperative follow-up period of ≤10 years, most patients recovered without evidence of disease recurrence (n = 18 [62.1%]). One patient had disease progression within 6 weeks and was alive with the disease after 13 months of follow-up.[22] Cases of permanent IX to XII cranial nerve palsy,[19,21] unilateral total deafness,[20] postoperative left palpebral ptosis, anisocoria with left myosis, and dysphonia[27] were also reported. Patients treated with radiotherapy and radionuclide therapy showed shrinkage or complete response to treatments, respectively.[28,37] The case left untreated resulted in sudden death after 1 year owing to rupture of the thoracic aorta and cardiac tamponade.[35]

DISCUSSION

CBTs are uncommon in the pediatric population and are often misdiagnosed as cervical lymphadenopathy or brachial cleft cysts, delaying timely surgical management.[25,33] The nonspecific location and silent growth of CBTs can obscure their clinical significance, causing clinicians to overlook the possibility of CBT in the differential diagnosis of pediatric cervical masses for years.[27,29,34] Interestingly, in at least two reported cases, the tumor presented during otologic examination[20,26] or with tinnitus or auditory symptoms.[21] In our case, the primary

clinical feature was a painful neck mass, with intermittent pain likely owing to intralesional nerve fiber involvement radiating to the neck. Given the atypical and sometimes silent presentation in children, CBTs should be considered in the differential diagnosis of cervical masses and cranial nerve compression symptoms.

When evaluating neck masses in the pediatric population, a comprehensive diagnostic approach is essential. Careful consideration for the risks of ionizing radiation in this age group, as opposed to adult patients with CBTs, is important. A thorough approach includes using a diagnostic pathway that includes ultrasound examination, possible computed tomography angiography, and magnetic resonance imaging. Additional tests like urinary metanephrines can confirm the diagnosis, but may sometimes be unremarkable, as in our case.[7] Owing to the high vascularity and proximity to large neurovascular structures, incisional biopsies are not generally recommended.[25] Angiographic studies can provide vital information about circulatory function and the need for arterial ligations or revascularization.[38] Assessing endocrine functionality is also of fundamental importance.[7]

Surgical resection is the primary treatment for CBTs.[7,22,39,40] Clinical observation only is not recommended generally owing to the risk of progressive growth and neurological deficits.[12] Surgery is challenging technically owing to the tumor's proximity to cranial nerves and the arterial system,[28] requiring an interdisciplinary perioperative management approach.[41] Angiographic assessment, including potential embolization, can help to prevent excessive intraoperative blood loss, which occurred in our case.[42] Neurological sequelae and cranial nerve injury have been reported in ≤44% of patients, especially in younger individuals.[7,25] For example, Hogan et al[32] described a young patient who experienced postoperative dysphagia. Such complications, although transient, pose significant postoperative care challenges. In our case, an NJ tube was required to manage nutritional difficulties associated with postoperative dysphagia, likely owing to hypoglossal nerve fibers injury.[7] Although less common, postoperative hypertension after CBT resection has been described as well. In our case, elevated blood pressure was detected in the postoperative care unit and persisted for 6 months after surgery. Although the pathophysiology of hypertension in our case cannot be ascertained fully, it has been previously described that the removal of CBTs exerting a significant mass effect may be associated with carotid baroreflex dysregulation.[43,44] Indeed, carotid body damage after adventitial stripping during CBT excision may lead to undesired hypertension with wide blood pressure variability.

Like in our case, Shamblin type III tumors may be challenging to dissect from the contiguous arteries and often require en bloc resection involving the external or internal carotid, or both, and thus may necessitate revascularization.[7,45] The decision to reconstruct vs ligate should be made on a case-by-case basis. Factors such as technical feasibility, the peculiar anatomy of the ICA, and its close proximity to the base of the skull should be taken into consideration. Additionally, a negative preoperative balloon occlusion test, despite not being 100% sensitive or specific, should be considered as well. In addition, vascular reconstruction after CBT resection has been associated with increased operative blood loss. However, it has not been shown to increase cranial nerve injury, hematoma formation, or stroke rates.[46]

Whereas surgical resection of CBTs is often curative, there have been some reports of recurrence after several years with metastatic dissemination.[47,48] Therefore, some authors have advocated the need for lifelong surveillance in these patients, consisting of yearly physical examinations; imaging of the head, neck, and abdomen regions; and plasma matrix metalloproteinases.[48]

Although CBTs are generally sporadic, early onset in children suggests a possible hereditary component.[5] In familial cases, genetic testing and counseling for both patients and their families must be considered.[45,49] In our case, genetic testing was offered to the patient's family, although they preferred not to undergo further evaluation. CBTs are also associated with chronic hypoxia, which is common in high-altitude residents or patients with chronic obstructive pulmonary or cyanotic heart disease.[39] These cases often present with cell hyperplasia at histological analysis.[50] Our patient was diagnosed with a case of sporadic CBT with hyperplasia; however, no correlation with chronic hypoxic phenomena was established as her asthma was under good therapeutic control, with no other apparent causes of gas exchange abnormalities.

Our case shared similarities and differences with other reported cases. Our patient, like most reported cases, was female and presented with a neck mass, the most common symptom. Unlike some patients, she did not exhibit cranial nerve compression symptoms before diagnosis.[7,16,21,22] Although some cases were bilateral or involved the glomus jugulare,[20,21,26,30] our patient had a single unilateral lesion. Although childhood CBT cases are often thought to be hereditary, the literature shows they are rarely familial, consistent with our patient's lack of family history.[21,26]

Preoperative embolization has been used by several authors,[7,23,24,26,29,32,36] with gross total resection being the mainstay of management.

Our review was limited significantly by the rare occurrence of CBT within the pediatric population. Moreover, pediatric and adult cases are presented together frequently, and extraction of individual data was not possible. These factors resulted in a small sample size of 30 reported cases, including ours. Also, the distribution of reported cases does not follow an even geographic distribution. Theories suggest that CBTs are associated

with high-altitude habitation, although altitude information was available only in two reported cases. Additionally, patients were presented from different socioeconomic environments, making timely diagnosis difficult in countries with limited access to modern imaging technologies. Future reports from diverse geographic backgrounds could enhance our understanding of pediatric CBT management strategies and possibly develop a clear diagnostic algorithm specific for this age group.

Despite advanced imaging methods facilitating CBT diagnosis and successful surgical treatment without sequelae in most cases, delayed diagnosis remains common. Given the metastatic potential of advanced or malignant CBTs and the fatal outcome of an untreated patient who was lost to follow-up, it is crucial to include CBTs in the differential diagnosis of neck masses in the pediatric population. Furthermore, the heterogenous clinical presentations described in the literature underscore the need for individualized care for each patient with CBT.

CONCLUSIONS

CBTs should always be included in the differential diagnosis of neck masses, although they are extremely uncommon as sporadic cases among the younger population. Despite the majority of lesions being benign, the potential for high surgical morbidity and vascularity emphasizes the need for meticulous perioperative and interdisciplinary management.

DISCLOSURES

None.

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