

Carotid body tumor in a nine-year-old patient

Dokuz yaşında bir hastada karotis cisim tümörü

Ahmet Baris Durukan^{1,2}, Hasan Alper Gurbuz^{1,3}

Institution where the research was done:
Liv Ankara Hospital, Ankara, Türkiye

Author Affiliations:

¹Department of Cardiovascular Surgery, Liv Ankara Hospital, Ankara, Türkiye

²Department of Cardiovascular Surgery, Istinye University, School of Medicine, Istanbul, Türkiye

³Department of Cardiovascular Surgery, Yüksek İhtisas University, School of Medicine, Ankara, Türkiye

ABSTRACT

Carotid body tumors are rarely encountered pathologies in the pediatric age group but still exist in the differential diagnosis of cervical painless masses. Genetic and familial background should be studied in addition to contrast imaging studies. Complete subadventitial resection, with or without prior embolization, which is still controversial, is the mainstay of therapy. Follow-up gains importance due to its nature. Herein, we report a nine-year-old girl presented with a unilateral asymptomatic mass on the neck with a diagnosis of carotid body tumor treated surgically without embolization. We emphasize the specific aspects of the pediatric age group.

Keywords: Carotid body tumor, operative, surgical procedures, therapeutic embolization.

Carotid body tumors, also known as glomus, are frequently located on the carotid bifurcation originating from its adventitial layer, are of neuroendocrine origin, and are the most common paragangliomas located on the head and neck region.^[1,2] The most common localization is the adrenal gland; only 3% involve the head and neck region, where they are mostly unilateral and solitary. The incidence varies between 1:30,000 and 1:100,000. Familial cases constitute 10% and arise in the pediatric age group.^[3] These are benign in nature but are considered malignant due to neighboring vital tissues and intracranial spread. They grow slowly, and diagnosis is by contrast enhancement

ÖZ

Karotis cisim tümörleri pediatrik yaş grubunda nadir görülen patolojilerdendir, ancak servikal ağrısız kitlelerin ayırıcı tanısında halen yer almaktadır. Kontrastlı görüntüleme çalışmalarına ek olarak genetik ve ailesel geçmiş de araştırılmalıdır. Halen tartışmalı olan önceden embolizasyon olsun ya da olmasın tam subadventisyal rezeksiyon tedavinin temel dayanağıdır. Takip, doğası gereği önem kazanır. Burada, boyunda tek taraflı asemptomatik kitle ile başvuran ve karotis cisim tümörü tanısıyla embolizasyon yapılmadan cerrahi olarak tedavi edilen dokuz yaşında bir kız çocuğu sunuldu. Pediatrik yaş grubunun belirli yönlerine vurgu yapıldı.

Anahtar sözcükler: Karotis cisim tümörü, operatif, cerrahi işlemler, terapötik embolizasyon.

in computed tomography and magnetic resonance imaging. Primary treatment is surgical resection with or without prior embolization, former easing resection by attenuating intraoperative bleeding.^[1] Herein, we report a case of a unilateral asymptomatic mass on the neck with a diagnosis of carotid body tumor treated surgically without embolization.

CASE REPORT

A nine-year-old female patient was admitted due to swelling on the right side of the neck. Physical examination revealed a painless 3×3 cm mass located medial to the middle one-third of the right sternocleidomastoid muscle. Carotid magnetic

Corresponding author: Ahmet Barış Durukan.

E-mail: barisdurukan@yahoo.com

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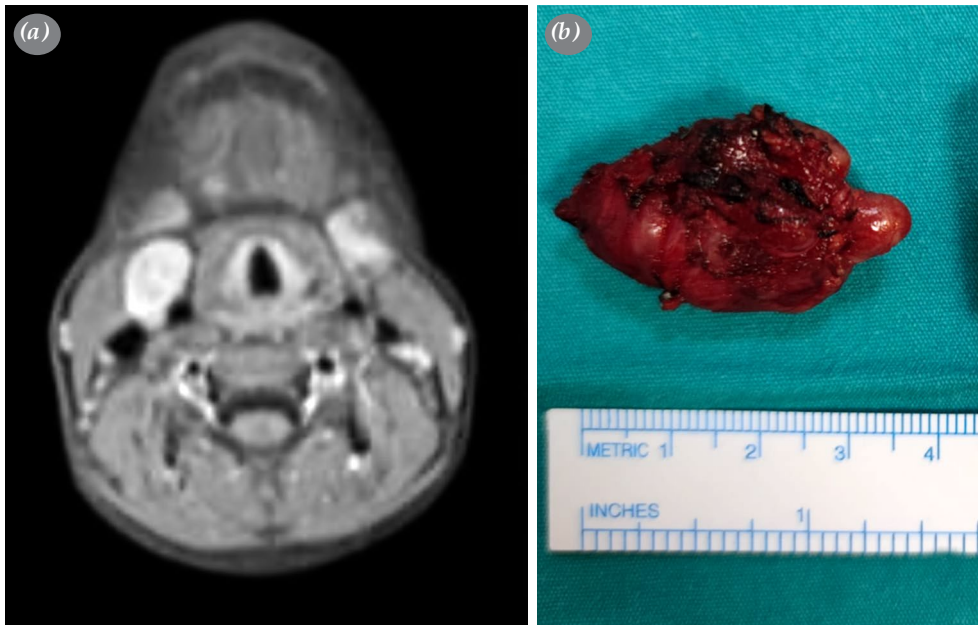


Figure 1. (a) A vascularized mass with a diameter of 4×4 cm is seen on magnetic resonance imaging angiography surrounding the right common, internal, and external carotid arteries. (b) Intraoperative image of the resected carotid body tumor.

resonance imaging angiography revealed a vascularized mass in the right carotid bifurcation 4×4 cm in size surrounding the right common, internal, and external carotid arteries (Figure 1a). Embolization was not performed due to the age of the patient and refusal of the procedure by the family due to possible risk of embolization. Under general anesthesia and near infrared spectroscopy monitoring, surgical resection was performed in the subadventitial plane (Figure 1b). No bleeding occurred, and neurologic complications did not develop perioperatively. The postoperative course was uneventful, and the patient was discharged on the second postoperative day. The pathological examination confirmed the diagnosis of a glomus tumor. No tumor recurrence was detected in the carotid computed tomography angiography at postoperative six months.

DISCUSSION

The gold-standard treatment of carotid body tumors is subadventitial resection, which may be challenging due to its relation with carotid arteries and neighboring cranial nerves.^[1-3] Hypoglossal, vagal, glossopharyngeal, and superior laryngeal nerves may be damaged leading to severe palsies. Stroke may occur during manipulation, and resection may be mortal or result in significant hemorrhage.^[2,3] Preoperative embolization ease resection, attenuate

intraoperative bleeding, and is recommended.^[4] Despite the advantages, pediatric age comprises a challenging patient group for embolization due to difficulty in procedure and increased risk of embolization.^[2] The family refused embolization procedure due to risk of cerebral embolization. Therefore, the surgery was performed without embolization, but neither hemorrhage nor any neurologic complication was observed by fine dissection and careful manipulation of the tissues.

The key point in treatment is that the procedure should be performed by experienced surgeons familiar with all surgical procedures involved in glomus tumor surgery. In the case we presented, the internal and external carotid arteries were almost completely wrapped with the tumor on the anterior aspect and the bulbous itself, but no invasion was observed on the arterial wall. It was a Shamblin class II tumor.^[5]

In the pediatric age group, cervical mass evaluation should include glomus tumors in the differential diagnosis, and the genetic basis should also be studied, particularly succinate dehydrogenase mutations, together with familial history examination. Pediatric patients have a genetic origin in almost half of cases.^[2] The patient we presented neither had a familial history, nor a genetic mutation. To the best of our knowledge, the youngest case reported was of an

eight-year-old patient,^[2] which we think makes this report scientifically valuable.

In conclusion, the pediatric age group makes up a specific group of glomus tumors. Genetic and familial background studies gain importance and shape the type of follow-up. Surgical resection is still the main treatment option, with debate over preoperative embolization.

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