PARAGANGLIOMA OF THE CAROTID BODY: CASE REPORT

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PARAGANGLIOMA OF THE CAROTID BODY: CASE REPORT (Abstract): With an incidence of 0.03% of all tumors, carotid body tumors (CBTs) are extremely rare. We present the case of a 63 year old female patient with an asymptomatic right sided neck mass of 3 cm diameter. Doppler ultrasound and contrast-enhanced CT confirmed the clinical suspicion of carotid body tumor. The patient underwent complete surgical excision of the Shamblin group II tumor and an adjacent lymph node, without preoperative embolization. The histopathological examination diagnosed a benign CBT. There were no postoperative complications at 1 and 6 months follow-ups. Keywords: PARAGANGLIOMA, CAROTID BODY TUMOR, SURGICAL TREATMENT.

Carotid body tumors (CBTs), also known as paragangliomas (PGLs) or glomus tumors, are very rare, with only a few vascular surgeons having encountered such pathology during their career. However, they are the most common paragangliomas of the head and neck (1) The very few studies on PGLs cited by the literature report contradicting male to female ratios (2-5), but agree the tumors are predominantly unilateral, with more than 50% on the right side of the neck. The mean age of presentation is between 49-55 years and recurrence rate is low (2, 3, 4).

The usual clinical presentation is an asymptomatic, slow-growing neck lump with Fontaine's sign (horizontally mobile, vertically fixed) (1, 2, 4, 5). In few cases, the CBT can cause various pressure symptoms: pain, cranial nerve palsy, dysphagia, odynophagia, hoarseness, dysphonia (1-4) or carotid sinus syndrome like symptoms: palpitations, fainting, falling (4, 5, 6).

Different imaging modalities are used preoperatively, most patients having multiple investigations. Doppler ultrasound (DUS) offers a non-invasive modality of diagnosing CBTs, which appear as solid, well-defined, hypervascular masses causing splaying of the carotid arteries. One series of patients reported CTA as the primary imaging modality, although most of those patients have been followed with DUS as well4. MRI is also effective for diagnosis, as is PET scan for tumors that are less than one cm in diameter (1). Historically, angiography has been advocated as mandatory for preoperative management of CBTs, as it
was shown to reduce the tumor mass and blood flow making surgical resection easier (3, 7, 8). More recent studies did not confirm the difference in blood loss and morbidity between embolized and non-embolized groups (9, 10).

CBTs can be classified as sporadic, familial or hyperplastic, with the former being the most frequent type. The familial type is encountered in only 20% of the cases and follows an autosomal dominant pattern of transmission (1, 19). Malignancy has been reported in less than 5-6% of all tumors (4, 11). It is generally accepted that malignancy is defined only by confirmation of metastasis in non-neuroectodermal tissue (12).

The recommended treatment of CBTs is complete surgical resection, with or without vessel reconstruction (1). Adjunctive therapy such as radiotherapy is reserved for metastatic disease, including patients with locally advanced extension that makes curative resection impossible. To aid pre-operative surgical planning, Shamblin introduced a classification scheme that divides CBTs in three groups according to size and local extension. First group consists of tumors that are small and easily removed. Group II includes tumors adherent to or partially surrounding vessels. Group III consists of tumors surrounding or encasing the vessels (13).

The classical surgical technique starts by ligation of the external carotid artery feeding branches, followed by dissection of the tumor from the carotid bifurcation towards the cranial side, in a sub adventitial plane. However, it has been shown that blood loss and cranial nerve injury can be improved with a modified technique that describes dissecting the tumor in a cranio-caudal direction, following the course of the ascending pharyngeal artery, suggested to be the tumor’s main blood supply (14).

Although the most frequent postoperative complication after CBT resection is cranial nerve palsy that resolves with time (15), a number of other complications have been described: uncontrollable hypertension, unilateral blindness (17) and persistent Horner’s syndrome (18). Mortality rates are between 0 and 3% and stroke rates between 0 and 8% (1).

Routine follow-up consists of clinical evaluation after 4-6 weeks and duplex ultrasound each year. CT or MRI is performed if new pathological findings occur on ultrasound (3).

**CASE REPORT**

A 63 years old female patient was admitted in our Vascular Surgery Department in May 2013 complaining of a 3 cm asymptomatic right sided neck mass. Her previous medical history included right sided breast cancer 35 years before, surgically treated with right mastectomy combined with axillaries clearance, and non-Hodgkin’s spleen lymphoma 6 years before, surgically treated with splenectomy followed by chemotherapy. At the time of admission, she was also being treated for high-risk grade III hypertension. As the patient was diagnosed with a sporadic CBT, no family screening was necessary.

Doppler ultrasound of the mass revealed a highly-vascularized tumor splaying the internal and external right carotid artery and also partially compressing the right internal jugular vein. A lymph node cranial to the tumor was also identified.

Contrast-enhanced CT of the neck and thorax revealed a 3 cm diameter mass located in the right carotid area, partially calcified in the lower segment, with intense
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annular contrast intake and post contrast hypo dense center, without evidence of extension to adjacent structures. The tumor had the same size compared to the preview CT scan performed 10 months earlier (fig. 1, 2).

![Native CT](image1)

**Fig. 1.** Native CT

![Contrast CT](image2)

**Fig. 2.** Contrast CT

Complete surgical removal of the tumor was performed under general anesthesia. The standard incision for carotid endarterectomy on the inner edge of the sternocleidomastoid muscle was undertaken. The common and internal carotid arteries were isolated with vessel loops, while the external carotid artery was clamped. Sub adventitial dissection of the tumor was then carried out from the carotid bifurcation in a cranial direction.

Excision of an adjacent lymph node completed the surgical procedure. The tumor was classified as belonging to Shamblin's group II (fig. 3, 4).

Histopathological examination of the excised tumor and lymph node, characterized the mass as a benign CBT. The lymph node had normal architecture and sinus histiocytosis (fig. 5, 6).

![Intraoperative image of the glomus tumor](image3)

**Fig. 3.** Intraoperative image of the glomus tumor

![Intraoperative appearance after the complete resection of the tumor](image4)

**Fig. 4.** Intraoperative appearance after the complete resection of the tumor
There were no immediate postoperative complications. The patient remained asymptomatic and complication free at clinical and Doppler ultrasound follow-up after one and six months postoperatively.

**DISCUSSION**

In patients presenting with swelling of the lateral neck, cranial nerve palsies, voice changes or auditory defects, a PGL of the neck should be included in the differential diagnosis. Careful evaluation of family history of tumors is important. Diagnosis is made on both clinical and imaging criteria. Our patient presented with a 3 cm asymptomatic right sided neck mass, highly suggestive for a carotid paraganglioma.

Imaging procedures, which include a mandatory ultrasound of soft tissue and vessels of the neck and CT or magnetic resonance angiogram (MRA) are recommended preoperatively. On ultrasound the tumor appears as a homogeneously hypoechoicogenic mass, while on Doppler scan a multidirectional hypervascularization is the main feature. Due to contrast medium enhancement the tumors give high signal intensity on MRA. The Shamblin groups can be predicted preoperatively on MRI.

CT can be useful to reveal relevant bony erosions or punctuate calcification in the tumor (20). To reduce radiation exposure and improve soft-tissue visualization, MRA is preferred to CT (20). DUS confirmed the clinical suspicion of a CBT for our patient and contrast-enhanced CT helped plan the surgical resection.

The clinical value of embolization is discussed controversially in the literature (10, 18, 21, 22). It can be considered in large tumors, because hypervascularization can be challenging during surgery. The risk for neurologic complications such as transient ischemic attacks (TIAs) or stroke caused by angiography or embolization is considerable. In large tumors surgical resection can be facilitated by preoperative embolization. Our patient had a Shamblin
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group II CBT, and therefore we decided the risks of embolization outweighed the benefits.

Complete surgical excision is the aim of any operative procedure for PGL and is the only curative option (22). Tumor doubling time is very slow and lies between 4 and 14 years; hence conservative treatment, such as a wait-and-see strategy, can also be discussed in elderly patients, who are free of PGL symptoms but do have other comorbidities. Ipsilateral lymphadenectomy should be performed simultaneously to identify malignant tumors. In these tumors malignancy is defined by metastases in non-neuroendocrine tissue (21). Complete resection of the tumor and excision of an adjacent lymph node was carried out in the case presented.

To reduce intra-operative bleeding, temporary clamping or ligation of the external carotid artery (ECA) may be required. Clamping but not ligation of the ECA was necessary during our surgical resection procedure, to minimize blood loss and improve cranial nerve detection. The larger the tumor the bigger the risk for inevitable nerve lesions or additional vascular procedures increases during surgery (4). Treatment in specialized vascular surgery centers is advisable because the incidence of vascular reconstructions is more than 25%.

Good blood pressure control is recommended. Baroreceptor failure syndrome (BFS) was described by Toma and Netterville and found in 16% of cases (23, 24). It might occur in patients after bilateral resection of the carotid glomus with postoperative acute hypertension and tachycardia. No complications were noted at 1 and 6 months postoperative follow-ups for our CBT patient.

Urinary vanillylmandelic acid and urinary or plasma metanephrine preoperative determination is important if an extra-adrenal tumor is suspected. Early diagnosis and surgical treatment of tumors before the tumor reaches 3 cm is recommended in order to avoid intra-operative nerve damage.

The patient must be referred to a surgeon who is familiar with the head and neck region and has the technical options for vascular reconstruction.

After histopathological confirmation of the paraganglioma, genetic testing and family examination is recommended.

REFERENCES


