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Diagnosis and treatment of carotid body paragangliomas: 20 years of experience

Raquel Carrilho Pichel

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INSTITUTO DE CIÊNCIAS BIOMÉDICAS ABEL SALAZAR
UNIVERSIDADE DO PORTO

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Diagnosis and treatment of carotid body paragangliomas: 20 years of experience

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Resumo

Introdução: Os paragangliomas do corpo carotídeo são tumores raros, mas constituem a forma mais comum de paragangliomas da cabeça e do pescoço. Estes são geralmente benignos e apresentam-se como uma massa cervical assintomática.

Objetivos: O objetivo deste estudo foi rever a abordagem dos doentes com tumores do corpo carotídeo tratados cirurgicamente no Serviço de Angiologia e Cirurgia Vascular deste Centro Hospitalar e os seus resultados.

Métodos: Foram revistos retrospetivamente os registos de 10 doentes acompanhados e tratados no Centro Hospitalar e Universitário do Porto com paragangliomas do corpo carotídeo entre 2000 e 2019. Foram analisados dados demográficos, características do tumor, abordagem diagnóstica, terapêutica e complicações. Adicionalmente foi conduzida uma pequena revisão da literatura sobre tumores do corpo carotídeo.

Resultados: Um total de 9 tumores do corpo carotídeo foram ressecados em 9 doentes. Destes, 5 eram mulheres (55.6%) e a idade média ao diagnóstico foi de 53.7 anos. Uma massa cervical não-dolorosa foi a queixa apresentada em 66.7%. A tomografia axial computadorizada foi utilizada em todos os casos para diagnóstico e 8 doentes foram ainda submetidos a outras modalidades de imagem. De acordo com a classificação de Shamblin, 5 eram do tipo II e 4 do tipo III. Foi confirmado que 1 tumor era funcional. Em todos os casos a ressecção cirúrgica foi bem-sucedida e não houve mortalidade peri-operatória. Complicações ocorreram no pós-operatório em 5 doentes, com lesões neurológicas permanentes em 2 (22.2%). Após um tempo médio de follow-up de 8.8 anos, não houve sinais de recorrência do tumor em nenhum doente ou mortalidade relacionada com o tratamento dos paragangliomas do corpo carotídeo.

Conclusões: A tumorectomia é o tratamento de escolha para os tumores do corpo carotídeo. Nesta pequena série de doentes com paragangliomas do corpo carotídeo, e de acordo com a literatura, os resultados foram positivos sem mortalidade ou recorrência associadas, mas com uma morbilidade considerável por lesões neurológicas permanentes em 22.2%

Palavras-chave: Carotid Body Tumor; Paraganglioma; Head and neck neoplasms.

Lista de Abreviaturas

CBT: carotid body tumors

CHUP: Centro Hospitalar Universitário do Porto

CNI: cranial nerve injury

CT: computed tomography

DSA: digital subtraction angiography

DUS: duplex ultrasound

FNAB: fine needle aspiration biopsy

ICD: International Classification of Diseases

MeSH: Medical Subject Headings

SDH: succinate dehydrogenase complex

TIA: transient ischemic attack

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Title Page

Title: Diagnosis and treatment of carotid body paragangliomas: 20 years of experience.

Original Article

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Declarations of interest: none

Abstract

Objectives: The purposes of this study were to review the management of patients with carotid body tumors treated surgically in the Angiology and Vascular Surgery Department of this center and evaluate their outcomes.

Design: Retrospective study on carotid body paragangliomas treated surgically in Hospital Santo António - Centro Hospitalar Universitário do Porto.

Materials and Methods: Records of 10 patients followed and treated in Hospital Santo António - Centro Hospitalar Universitário do Porto for paragangliomas of the carotid body between 2000 and 2019 were retrospectively reviewed. Demographic data, lesion characteristics, diagnostic approach, therapeutic and outcomes were analyzed. A small revision of the literature on carotid body paragangliomas was conducted as well.

Results: A total of 9 carotid body tumors were resected in 9 patients. Five were women (55.6%) and the mean age at diagnosis was 53.7 years. A nontender neck mass was the presenting complaint in 66.7%. Computed tomography scan was used in all cases for diagnosis and 8 patients had additional imaging modalities. According to the Shamblin classification, 5 were type II and 4 type III. Only 1 tumor was confirmed functional. In all cases surgical resection was successful and there was no perioperative mortality. Complications occurred postoperatively in 5 patients, with cranial nerve injury remaining permanent in 2 (22.2%). After a mean follow-up time of 8.8 years there were no signs of tumor recurrence in any patient or mortality related to carotid body paragangliomas treatment.

Conclusions: Surgical resection is the treatment of choice for carotid body tumors. In this small series of patients with paragangliomas of the carotid body and in accordance to the literature the outcomes were positive with no mortality or recurrence associated but with a noteworthy morbidity, as cranial nerve injury was permanent in 22.2 %

KEY WORDS: Carotid Body Tumor; Paraganglioma; Head and neck neoplasms.

Introduction

Paragangliomas are neural crest tumors located along the paravertebral sympathetic and parasympathetic chains. The head and neck region is a common site for paragangliomas accounting for up to 70% of extra-adrenal paragangliomas.[1]

Although carotid body paragangliomas, or carotid body tumors (CBT), are rare (1:30 000) they constitute the majority of head and neck neuroectoermal tumors and their approach still presents a challenge for vascular surgeons.[2]

There are three recognized etiological types of CBT: sporadic, hyperplasic and familiar. The sporadic form is the most frequent and the hyperplasic tumors have been correlated with chronic hypoxia contexts.

Regarding the family form, which accounts for up to 30% of the cases, the transmission is autosomal dominant with variable penetrance and is characterized by the frequency of multifocal localization as high as 80%. Multiple germline mutations have been identified and the mutation of genes associated with the mitochondrial succinate dehydrogenase complex (SDH) involved in the cycle of Krebs, especially the SDH subunit D gene, which is the most frequent.[3] [4]

Clinically, the tumors present as an indolent, mobile and painless lateral neck mass located anteriorly to the sterno-cleido-mastoid muscle at the level of the hyoid bone and may pulsate. Symptoms of direct involvement or dysfunction of cranial nerves such as cranial nerve palsies, voice changes or auditory defects may also be present. [5]

These lesions were first described as benign, slow-growing neoplasms. However, malignancy, defined as the presence of metastases, as there are no clear histologic characteristics, has been reported in less than 10% of the cases. [6]

The diagnosis of carotid body paragangliomas is based on clinical history and imaging findings, such as duplex ultrasound (DUS), computed tomography (CT), magnetic resonance angiography and digital subtraction angiography (DSA). DSA is the gold standard in this highly vascularized lesion. It demonstrates tumor blood supply and widening of the carotid bifurcation by a well-defined tumor blush, which is a classic pathognomonic finding. The assessment of the tumor vascular supplying is fundamental for preoperative embolization.[7] If the diagnosis of CBT is suspected, biopsy is contraindicated, as the risk of hemorrhage is significant.[8]

Since Shamblin et al. proposed the classification of CBT based on the size and involvement of carotid vessels in 1971[9], it has been widely used as a predictor of intra-operative technical difficulties (Table I). The size of tumour is positively correlated with the Shamblin classification because the paragangliomas become more adherent to carotid vessels as they enlarge, and

consequently appears to be associated with an increased incidence of major peri-operative adverse events, stroke and cranial nerve injury (CNI).[10] More recently, new predictors of complications of CBT resection have been proposed. Smaller distance of the tumor to the base of the skull has been associated with higher CNI and blood loss.[10]

The treatment of carotid body paragangliomas englobes endovascular preoperative techniques, surgical resection, radiotherapy and vigilance. Surgery has been considered the gold standard in selected patients to prevent local progression and malignancy but in selected cases radiotherapy or a watchful waiting policy is preferred.[5]

Pre-operative embolization clinical value remains controversial although it allows pre-operative tumor size reduction and helps to reduce intraoperative blood loss. Timing and inherent complications are still matter of debate. [11, 12]

The detection of the tumor in an early stage improves surgical treatment outcomes but the morbidity and mortality of the CBT resection is not trivial, especially with perioperative vascular complications and cranial nerve deficits. After surgery, these patients require follow-up and regular imaging examinations due to the possibility of recurrence or occurrence of multiple tumors. [5]

The aim of this study was to review the methodology of CBT diagnosis and treatment in this center and compare the results with the international literature

Materials and Methods

A literature search was undertaken identifying reviews and case reports published between January 2000 and December 2019 through the PubMed/MEDLINE databases. The key words were “Carotid Body Tumor” and “Paraganglioma” from the Medical Subject Headings (MeSH) Index Medicus. References quoted in useful articles were also looked independently of the year of publication when relevant.

Medical records of patients with CBT treated between 2000 and 2019 in our center, Hospital de Santo António, Centro Hospitalar Universitário do Porto (CHUP), were retrospectively analyzed. The patients undergoing treatment for carotid body paraganglioma were identified using a search for the associated International Classification of Diseases (ICD)-9 and ICD-10 diagnosis code. These records were reviewed for demographic, etiologic, diagnostic, and therapeutic features. The extracted variables were gender, age at time of diagnosis, symptoms, bilateral involvement, family history, diagnostic methods, tumor characteristics, presence of genetic mutations, treatment modality, surgical complications and outcomes.

The tumors were divided using the Shamblin classification based on the imaging and operative findings.

For statistical analysis IBM SPSS Statistics Version 26 was used and the results were compiled and analyzed using descriptive statistics.

The institutional review board and local Ethics Commission authorized this study, and waived the need of patient consent.

Results

A total of 10 cases were identified as a corresponding match for diagnose of CBT, treated surgically in CHUP between January 1, 2000 and December 31, 2019. One patient was excluded due to insufficient data. All 9 CBT were surgically resected by tumorectomy and were histologically proven CBT. There were 5 female patients (55.6%) and the mean age at time of diagnosis was 53.7 years (range, 34-79 years). Concerning etiology, 7 patients had the sporadic form and 2 had family history of CBT. There were no bilateral tumors or malignant paragangliomas.

A painless neck mass was the most common presentation, and as a main symptom in 6 patients (66.7%). One complained of odynophagia alongside. Two patients had symptoms of increased catecholamines (hypertension, palpitations or flushes) and the CBT secretory function was assessed confirming catecholamine-secretion in one case. Preoperative adrenergic blockade was given to that patient. Two cases were asymptomatic, as CBT was an incidental finding on imaging modalities. At initial presentation, there were no symptoms of cranial nerve deficit.

Shamblin classification was assigned according to preoperative imaging, operative findings and pathologic confirmation of the tumor relation to the blood vessels but it did not show any significant influence either on the presentation or on complications. There were 4 Shamblin II and 5 Shamblin III tumors.

There were no signs of metastatic disease at presentation or during follow-up. The diagnostic modalities used in the diagnosis of the 9 CBT are listed in table II. CT scan was the most common imaging tool, used in all 9 patients (4 CT angiography). Eight patients underwent more than one diagnostic imaging modality, including also DUS (n=4), ultrasound (n=4) and angiography (n=3). Two patients underwent fine needle aspiration biopsy (FNAB), with no associated complications.

Despite the familial history, present in 2 patients, the SDH germline mutations were not searched in any case.

All patients were managed by CBT total tumorectomy. The surgical technique included precise anatomic dissection and vascular control prior to attempted tumor excision. The dissection to remove the CBT was carried out along the arterial subadventitial plane to allow for complete local tumor excision, as well as preservation of critical vascular structures. The craniocaudal approach was used. In none of the patients vascular resection and reconstruction was described.

Preoperative embolization with embolizing microparticles was done in 8 CBT. Criteria were provably size tumor and involvement of main vessels. No strokes or other major complications occurred after pre-embolization

All 9 CBT resections were technically successful, and no perioperative mortality befell. Complications occurred in 5 patients and are summarized in Table II. The most common postoperative complication was temporary cranial nerve deficit – as labial commissure deviation (n=1) and pain or difficulty on deglutition (n=2). Other postoperative complications included hoarseness (n=1) and headache (n=1), both remaining permanently, and temporary scar hypoesthesia (n=1). No other complications were identified.

No further radiotherapy treatments were necessary as the excisions were complete.

The median follow-up time was 8.8 ± 5.9 years and during that time there was no tumor recurrence or delayed complications from the CBT surgery.

Discussion

CBT are usually benign slow growing vascular head and neck neoplasms and can remain asymptomatic for many years. They are very challenging neck tumor due to their unpredictable course, malignant potential and complications associated with larger tumors. Early and complete surgical resection is the recommended treatment. This is a report of the CHUP Angiology and Vascular Surgery Department series of patients with paragangliomas of the carotid body.

The majority of CBT are sporadic, with a mean age of presentation ranging from 35.4 to 56.6 years.[13, 14] In our series the mean age was 53.7 years and the female to male ratio was 1.25:1, which is consistent with the slight female prevalence described in the literature.[2, 6, 15, 16] The incidence of familial CBT in our series (22.2%) is concordant with other series.[13, 16] No bilateral tumors were found in our study but CBT have been reported bilateral in 5% of sporadic cases and 33% in familial cases.[6] The familial form is associated with SDH germline mutations. Regarding head and neck paragangliomas the most prevalent are SDH genes, with SDHD mutations representing approximately 80%, followed by the subunit B mutations at around 20% and the remainder subunit C mutations.[4] Fruhmann et al reinforced that careful evaluation of family history regarding tumors is important and recommend genetic testing for all paragangliomas because individuals with germline mutations may also present as clinically sporadic paragangliomas.[17] No report of family screening is referred in patients' records.

Most CBT are non secreting tumors, but functional CBT have been describe in up to 18% of the cases and can lead to presenting symptoms triggered by the catecholamine secretion.[14] Our series included a patient with functional CBT (11.1%) who presented an episode of increased catecholamines alongside a painless neck mass as the main complaints that led to diagnose.

In agreement with other authors the most common clinical presentation in this series was a painless slow growing neck mass. In a recent meta-analysis by Robertson et al. a neck mass was the symptom of CBT in 75.3% of the cases in the 92 studies reviewed.[18] Therefore, paraganglioma diagnosis should be considered in every patient with an indolent neck mass. FNAB should not be done because of high risk of bleeding or damaging near noble neurovascular structures. In our set 2 patients underwent this procedure without complications, still it does not bring any advantage, as the cytology results cannot differentiate benign from malign lesions.

For CBT diagnosis, a thorough history and physical examination should be performed including cranial nerves examination. Imaging investigations aim to confirm the diagnosis, evaluate the size of the lesion and allow proper operative planning. The contralateral side must also be evaluated for CBT. In our patients DUS, CT scan and CT angiography were the preferred

imaging techniques. Despite DUS been able to identify a highly vascularized tumor and widening of carotid bifurcation additional investigation with CT helps identifying the dimensions and anatomical correlations of the tumor. Angiography allows assessment of the vessels supplying the tumor and preoperative embolization. Nevertheless, ultrasound may be used for screening because it is non invasive and widely available. [5]

As for malignity, no metastatic disease was identified in the CBT excised in our center, so this percentage is compatible with the lowest value in the range of malignity described in the literature - 0 to 11.1%. [2, 5, 6, 19, 20] No recurrence was detected in this series but a lifelong surveillance based on physical examination and imaging techniques, if necessary, seems to be indicated.

In an attempt to decrease the morbidity associated with large lesions, particularly Shamblin III tumors, preoperative embolization as been preconized by some authors, by selectively approach the tumor irrigation.[21] This procedure has its one inherent risk and the results in the literature are not consensual. [12, 22] In our series 8 patients (88.9%), 4 Shamblin II and 4 Shamblin III, had arterial embolization prior to surgery and there were no complications associated with the procedure but no significative differences in terms of morbidity compared with the patients who did not receive pre-embolization in same Shamblin class tumors could be weighed.

As mentioned above, surgical resection is the treatment of choice for CBT but it is associated with significant morbidly and mortality. As described in a recent review and meta-analysis by Robertson et al, there is a 2.2% 30-day mortality and 3.5% 30-day stroke associated with CBT resection.[18] During the procedures, all neurovascular structures should be identified to reduce complications. Cerebrovascular complications such as stroke or transient ischemic attack (TIA) associated with or without manipulation of major neck vessels can occur, and incidence has been reported to be 0 to 18.1%. [5, 6, 13, 19, 23] Postoperative CNI is common, and incidence of CN deficits is higher in Shamblin III tumors but most are temporary. Permanent deficits occur in 11.2%.[18] In our series there were no cerebrovascular morbidity and from the 5 patient who had postoperative complication only 2 (22.2%) remained with permanent hoarseness and headache.

As most publications on CBT, this is a single center experience review, encountering only few cases which makes it difficult to interpret the results (Table III). However, the experience from this casuistry shows that the recommended surgical excision is associated with long-term positive outcomes, such as very good survival and low recurrence rates, but also with a meaningful risk of CNI, which could cause great impact in patients' life when permanent. Preoperative embolization

on the day preceding surgery reduces tumour size enhancing the possibility of total tumour resection, reducing significantly peri and postoperative complications.

The limitations of this study are associated with its retrospective character, specially when it came to identifying the patients with CBT diagnose which depended from the ICD codification, and a huge lack of data in most of the medical records, specially before 2015.

References

1. Wieneke JA, Smith A. Paraganglioma: carotid body tumor. Head Neck Pathol. 2009;3(4):303-6.
2. Sajid MS, Hamilton G, Baker DM, Joint Vascular Research G. A multicenter review of carotid body tumour management. Eur J Vasc Endovasc Surg. 2007;34(2):127-30.
3. Ding Y, Feng Y, Wells M, Huang Z, Chen X. SDHx gene detection and clinical Phenotypic analysis of multiple paraganglioma in the head and neck. Laryngoscope. 2019;129(2):E67-E71.
4. Williams MD, Tischler AS. Update from the 4th Edition of the World Health Organization Classification of Head and Neck Tumours: Paragangliomas. Head Neck Pathol. 2017;11(1):88-95.
5. Dorobisz K, Dorobisz T, Temporale H, Zatonski T, Kubacka M, Chabowski M, et al. Diagnostic and Therapeutic Difficulties in Carotid Body Paragangliomas, Based on Clinical Experience and a Review of the Literature. Adv Clin Exp Med. 2016;25(6):1173-7.
6. Gad A, Sayed A, Elwan H, Fouad FM, Kamal Eldin H, Khairy H, et al. Carotid body tumors: a review of 25 years experience in diagnosis and management of 56 tumors. Ann Vasc Dis. 2014;7(3):292-9.
7. Singh D, Pinjala RK, Reddy RC, Satya Vani PV. Management for carotid body paragangliomas. Interact Cardiovasc Thorac Surg. 2006;5(6):692-5.
8. Williams MD. Paragangliomas of the Head and Neck: An Overview from Diagnosis to Genetics. Head Neck Pathol. 2017;11(3):278-87.
9. Shamblin WR, ReMine WH, Sheps SG, Harrison EG, Jr. Carotid body tumor (chemodectoma). Clinicopathologic analysis of ninety cases. Am J Surg. 1971;122(6):732-9.
10. Kim GY, Lawrence PF, Moridzadeh RS, Zimmerman K, Munoz A, Luna-Ortiz K, et al. New predictors of complications in carotid body tumor resection. J Vasc Surg. 2017;65(6):1673-9.
11. Power AH, Bower TC, Kasperbauer J, Link MJ, Oderich G, Cloft H, et al. Impact of preoperative embolization on outcomes of carotid body tumor resections. J Vasc Surg. 2012;56(4):979-89.
12. Jackson RS, Myhill JA, Padhya TA, McCaffrey JC, McCaffrey TV, Mhaskar RS. The Effects of Preoperative Embolization on Carotid Body Paraganglioma Surgery: A Systematic Review and Meta-analysis. Otolaryngol Head Neck Surg. 2015;153(6):943-50.
13. Darouassi Y, Alaoui M, Mliha Touati M, Al Maghraoui O, En-Nouali A, Bouaity B, et al. Carotid Body Tumors: A Case Series and Review of the Literature. Ann Vasc Surg. 2017;43:265-71.
14. Dixon JL, Atkins MD, Bohannon WT, Buckley CJ, Lairmore TC. Surgical management of carotid body tumors: a 15-year single institution experience employing an interdisciplinary approach. Proc (Bayl Univ Med Cent). 2016;29(1):16-20.
15. Metheetrairut C, Chotikavanich C, Keskool P, Suphaphongs N. Carotid body tumor: a 25-year experience. Eur Arch Otorhinolaryngol. 2016;273(8):2171-9.
16. Davila VJ, Chang JM, Stone WM, Fowl RJ, Bower TC, Hinni ML, et al. Current surgical management of carotid body tumors. J Vasc Surg. 2016;64(6):1703-10.
17. Fruhmann J, Geigl JB, Konstantiniuk P, Cohnert TU. Paraganglioma of the carotid body: treatment strategy and SDH-gene mutations. Eur J Vasc Endovasc Surg. 2013;45(5):431-6.
18. Robertson V, Poli F, Hobson B, Saratzis A, Ross Naylor A. A Systematic Review and Meta-Analysis of the Presentation and Surgical Management of Patients With Carotid Body Tumours. Eur J Vasc Endovasc Surg. 2019;57(4):477-86.
19. Lim JY, Kim J, Kim SH, Lee S, Lim YC, Kim JW, et al. Surgical treatment of carotid body paragangliomas: outcomes and complications according to the shamblin classification. Clin Exp Otorhinolaryngol. 2010;3(2):91-5.
20. Kohler HF, Carvalho AL, Mattos Granja NV, Nishinari K, Kowalski LP. Surgical treatment of paragangliomas of the carotid bifurcation: results of 36 patients. Head Neck. 2004;26(12):1058-63.

21. Li J, Wang S, Zee C, Yang J, Chen W, Zhuang W, et al. Preoperative angiography and transarterial embolization in the management of carotid body tumor: a single-center, 10-year experience. Neurosurgery. 2010;67(4):941-8; discussion 8.
22. Abu-Ghanem S, Yehuda M, Carmel NN, Abergel A, Fliss DM. Impact of preoperative embolization on the outcomes of carotid body tumor surgery: A meta-analysis and review of the literature. Head Neck. 2016;38 Suppl 1:E2386-94.
23. Valentim H, Goncalves F, Vasconcelos L, Garcia A, Emilia Ferreira M, Albuquerque ECJ, et al. [Carotid body tumors. A 10-years experience in the management of the disease]. Rev Port Cir Cardiotorac Vasc. 2008;15(3):145-9.
24. Casarim AL, Tincani AJ, Del Negro A, Aguiar CG, Fanni RV, Martins AS. Carotid body tumor: retrospective analysis on 22 patients. Sao Paulo Med J. 2014;132(3):133-9.
25. Lamblin E, Atallah I, Reyt E, Schmerber S, Magne JL, Righini CA. Neurovascular complications following carotid body paraganglioma resection. Eur Ann Otorhinolaryngol Head Neck Dis. 2016;133(5):319-24.
26. Kotelis D, Rizos T, Geisbusch P, Attigah N, Ringleb P, Hacke W, et al. Late outcome after surgical management of carotid body tumors from a 20-year single-center experience. Langenbecks Arch Surg. 2009;394(2):339-44.
27. Law Y, Chan YC, Cheng SW. Surgical management of carotid body tumor - Is Shamblin classification sufficient to predict surgical outcome? Vascular. 2017;25(2):184-9.
28. Amato B, Bianco T, Compagna R, Siano M, Esposito G, Buffone G, et al. Surgical resection of carotid body paragangliomas: 10 years of experience. Am J Surg. 2014;207(2):293-8.

Tables

Table I – Shamblin classification of carotid body tumors

Class	Tumor characteristics
I	Relatively small, minimally attachment to the carotid vessels; complete resection usually without difficulty.
II	Moderately attachment to the internal and external carotid artery; complete resection more challenging.
III	Encase the carotid vessels; complete resection often requires major vessels reconstruction.

Table II –Characteristics of 9 cases of carotid body tumors

Patient	Age	Gender	Family history	Presenting symptoms	Diagnosis modalities	Tumor size (cm)	Shamblin class	Functional tumor	Preop embolization	Postop complications	Mortality related to CBT	Follow-up (years)	Recurrence
1	69	M	-	Incidental finding	CT scan; angiography	---	II	-	+	Long-term (>1year postoperative) hoarseness	-	19	-
2	46	F	-	Increased catecholamines	CT scan; DUS	---	III	-	+	-	-	15	-
3	73	F	-	Painless neck mass	CT scan; DUS; angiography	---	II	-	+	-	-	13	-
4	79	M	-	Incidental finding	CT angiography; DUS	---	III	-	+	-	-	9	-
5	34	M	-	Odynophagia; neck mass	CT scan; ultrasound; FNAB	---	II	-	+	Temporary scar hypoesthesia	-	7	-
6	51	F	-	Painless neck mass; palpitations	CT angiography; ultrasound; FNAB	3	II	+	+	-	-	7	-
7	45	M	-	Painless neck mass	CT scan; ultrasound; DUS; angiography	4	III	-	+	Temporary pain on deglutition	-	5	-
8	40	F	+	Painless neck mass	CT angiography	3	II	-	-	Long-term (>1year postoperative) headache	-	3	-
9	46	F	+	Painless neck mass	CT angiography; ultrasound	4	III	-	+	Temporary labial commissure deviation and difficulty on deglutition	-	1	-

Table III - Demographic features and outcomes data literature review

Study	Patients (N)	F:M ratio	Mean age presentation (years)	Malignant	Bilateral	CNI - permanent	Stroke/ TIA	Mortality - related to CBT
Sajid et al. 2007[2]	95	1.9:1	55	4.2%	17%	1%	1%	1%
Dorobisz et al. 2016[5]	47	1: 1.5	45	0%	4.2%	10.6%	6%	2.1%
Dixon et al. 2016[14]	17	4.7:1	56.6	0%	29%	0%	5.8%	0%
Casarim et al. 2014[24]	22	1.75:1	43	---	9%	13.6%	4.5%	0%
Gad et al. 2014[6]	54	2.3:1	42	0%	3.6%	3.57%	18.1%	0%
Lim et al. 2010[19]	13	1.17:1	44	0%	15%	23%	15.4%	7%
Valentim et al. 2008[23]	8	3:1	53	0%	0%	12.5%	0%	0%
Metheetrairut et al. 2016[15]	38	1.92:1	36.9	0%	18.4%	20%	5%	0%
Kohler et al. 2004[20]	26	2.6:1	39	15.4%	---	15.4%	0%	0%
Lamblin et al. 2016[25]	49	1.2:1	49	4%	18.4%	17%	6.1%	2%
Kotelis et al. 2009[26]	17	1.83:1	49	0%	0%	11.8%	0%	5.6%
Davila et al. 2016[16]	183	2:1	49	0%	8.7%	5.5%	1%	0.55%
Darouassi et al. 2017[13]	10	2.33:1	35.4	10%	10%	0%	0%	0%
Law et al. 2017[27]	20	4:1	36	5%	5%	14%	10%	0%
Amato et al. 2014[28]	31	1.58:1	48	0%	9.7%	12.9%	6%	3%
Our series Pichel et al. 2020	9	1.25:1	53.7	0%	0%	22.2%	0%	0%