An evaluation on multidisciplinary management of carotid body paragangliomas: a report of seven cases

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Abstract
Carotid body paragangliomas (CBPGLs) are a rare neoplasms of the neuroendocrine system that affect the carotid glomus. The aim of this study is to improve their management in our Departments. This retrospective analysis reports family history, clinical presentation, imaging diagnostics, Shamblin classification, surgical treatment, complications, and the outcome of seven patients with CBPGLs. All lesions were represented by a painless cervical mass, with no functional or bilateral neck tumors. One patient had two different localizations (the second one was a glomus tumor of the right prelachrymal sac), and a family history for CBPGL. All neck tumors were diagnosed during duplex ultrasound corroborated by magnetic resonance imaging (MRI), and by magnetic resonance angiography (MR-A). They presented a diameter between 3 and 5 cm (MRI). Complete subadventitial resection of the tumor was performed in all patients, with no preoperative embolization in any of the cases. The CBPGLs were confirmed on histopathology and immunohistochemistry. Lymph node metastasis was not found in any of the cases. Mortality and perioperative stroke rates were null. Transitory cranial nerve deficit occurred in one case without permanent palsy. After a follow-up of three years in each patient, there were no signs of tumor recurrence in any of the cases. Relatively early diagnosis of CBPGL was possible in our seven patients using multidisciplinary management. Preoperative planning of the surgical procedure by integrated diagnostic imaging was essential in our study to operate only Shamblin group II tumors, minimizing the known risk of complications associated with large CBPGL (group III).

Keywords: carotid body paragangliomas, duplex ultrasonography, resection.

Introduction
Macroscopically, the carotid body is a well circumscribed, round, reddish-brown highly specialized organ. Its measurements are approximately 5×3×2 mm and it is located in the adventitia of the carotid bifurcation [1]. Its feeding vessels run primarily from the ascending pharyngeal artery (a branch of external carotid artery – ECA), but may receive branches from the internal carotid artery (ICA) bulb, and innervated through the IX (glossopharyngeal) and X (vagus) nerves [2–5]. Microscopically, the tumors are highly vascular; between the many capillaries are clusters of cells, including supporting cells and chief cells [1]. Cytochemical techniques usually demonstrate epinephrine, norepinephrine and serotonin in these cells [1]. The function of the carotid body is related to the autonomous control of the respiratory and cardiovascular systems, as well as blood temperature [3–5]. It is a chemoreceptor organ that is stimulated by hypercapnea, hypoxia, and acidosis, which controls the autonomous control drive by increasing the sympathetic flow [3–5].

Carotid body paragangliomas (CBPGLs) are relatively rare tumors arising from paraganglionic cells of the carotid body, which develop from both mesodermal elements of the third branchial arch and neural elements originating from the neural crest ectoderm [2, 3, 6–8]. Van Haller, in 1743, first described the carotid body while the term “paraganglia” was introduced by Kohn in the early XXth century [1, 9].

The main purpose of our study is to establish a better-standardized proceeding in the diagnosis (clinical and imaging characteristics) and surgical treatment of these tumors, in our Departments in order to provide the best outcome of such patients.

Case presentations
This is a retrospective analysis of the medical records of all cases diagnosed with CBPGLs and operated on in our Departments between March 2009 and December 2012. There were seven patients (five women, two men with mean age of 54.7 years, with an overall age ranging from 41 to 69 years). All the preoperative, operative and postoperative data were studied for each case and the follow-up by the medical records. All patients were given a medical history questionnaire. Familial disease was
initially determined by pedigree analysis. All cases underwent a complete head and neck examination, through clinical neurological, ear nose throat (ENT: preoperative and postoperative laryngoscopy as well as phoniatric evaluation), surgical, anesthesiologist, and ophthalmological practices. The clinical characteristics of the CBPGLs were represented by: site, size, consistency, and pulsatility of the neck mass, and the possible preoperative complications (craniofacial involvement, etc.). All patients were examined for possible functional tumors (typical symptoms such as flushing, palpitations, or headaches). Before surgery, all cases with a presumed CBPGL underwent combined imaging techniques in order to classify the type of tumor and plan the proper treatment. All patients have been evaluated by the first author of this study, by duplex ultrasound as first step (with a 7.5–10 MHz linear array transducer, combining B mode and color Doppler/pulsed-wave Doppler ultrasound).

All cases underwent a second examination, represented by magnetic resonance imaging – MRI (1.5 T), and magnetic resonance angiography (MR-A). The characteristic findings on MRI and MR-A were studied for lesion shape, margin, signal intensity, angle of common carotid artery (CCA) bifurcation, and the relationship between the great vessels and the carotid space mass. All lesions were classified using the MRI measurements into three groups according to the latero-lateral diameter of the tumor: group I (<3 cm), group II (3–5 cm), and group III (>5 cm).

All patients were operated by the same team (one ENT and one surgeon). We analyzed surgical treatment modalities used. All interventions were performed under general anesthesia. The tumors were confirmed on histopathology and immunohistochemistry, and were screened for malignancy. The main criteria of malignancy were obtained from operative notes, e.g., lymph node metastases and aggressive local infiltration of adjacent tissues.

We examined surgical complications and outcome. Major outcomes included peri-operative mortality, stroke/transient ischemic attack (TIA), and cranial nerve injury as well as recurrences. All patients were followed-up clinically every three months for the first year, and twice/year for the next two years, with yearly duplex ultrasound.

Clinical presentation

All unilateral CBPGLs were discovered upon patient self examination. The main clinical data are shown in Table 1.

Table 1. Clinical presentation of unilateral carotid body paragangliomas (CBPGLs)

<table>
<thead>
<tr>
<th>No.</th>
<th>Site/size (cm) of the neck mass (MRI)</th>
<th>Consistency of the neck mass</th>
<th>Pulsatility of the neck mass</th>
<th>Clinically functional tumor</th>
<th>Preoperative local complications</th>
<th>Shamblin classification</th>
<th>Post-operative local complications</th>
<th>Post-operative TIA/stroke</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Right/4.1</td>
<td>Painless</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Stage II</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>2.</td>
<td>Left/4.3</td>
<td>Painless</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Stage II</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>3.</td>
<td>Left/4.2</td>
<td>Painless</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Stage II</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>4.</td>
<td>Right/4.2</td>
<td>Painless</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Stage II</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>5.</td>
<td>Left/4.4</td>
<td>Painless</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Stage II</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>6.</td>
<td>Right/4.1</td>
<td>Painless</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Stage II</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>7.</td>
<td>Left/4.7</td>
<td>Painless</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Stage II</td>
<td>Yes</td>
<td>No</td>
</tr>
</tbody>
</table>

MRI: Magnetic resonance imaging; TIA: Transient ischemic attack.

Physical examination revealed in all seven cases a unilateral painless palpable pulsatile neck mass, laterally mobile but vertically fixed, located just anterior to the sternocleidomastoid muscle at the level of the hyoid bone, and below the angle of the mandible. The neck masses were firm and incompressible, and were suspected of the tumor due to the transmitted pulsation. There was no evidence of clinically functioning tumors (with none of the patients having signs of increased catecholamine secretion: flushing, palpitations), and no bilateral involvement of carotid bifurcation in any of the cases.

No neurological abnormalities caused by vagal or hypoglossal nerve involvement (dysphagia/dysphonia), or by cervical sympathetic nerve impingement (Horner’s syndrome) were reported before surgical excision.

The first patient had a family history for CBPGL, and two different localizations (the second one was a glomus tumor of the right prelachrymal sac).

Imaging diagnostics

All patients have been evaluated by duplex ultrasound as the first step, followed by a second group of examinations, represented by MRI and MR-A.

Duplex ultrasound characteristics

The topographic relation of the tumors to the carotid arteries and the internal jugular vein and the patterns of vascularization were assessed in all seven cases:

(a) B-scan imaging revealed in all cases a well-defined, solid, inhomogeneous, hypoechoic mass, located at the carotid bifurcation that pushed the carotid arteries apart.

(b) Pulsed Doppler analysis of blood flows indicated in all tumors low-resistance flow patterns [with a low resistance index (RI), and a high diastolic component], obtained from multiple sites within the mass of the tumor. An arterio-venous shunt could be seen in all cases, with accelerate and turbulent flow in the vessels of the tumor.

(c) The color Doppler imaging revealed in all patients a characteristic broadening (wide splaying) of the carotid bifurcation by a hyper-vascular mass (the highly vascularized tumor), with shifting of both the ICA and the internal jugular vein posterior and laterally and of the ECA anterior and medially. The hypervascularity of the CBPGLs appeared in all these cases as an abundant flow signal (irregular color signals) with flow direction being predominantly upward (according to the direction of tumor growth and vascular supply). All seven carotid body tumors could be assessed to their full extent (Figure 1).
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Figure 1 – Duplex ultrasound – longitudinal view: carotid body tumor at right carotid bifurcation. Hypervascular cervical mass widening the right carotid bifurcation (the internal and external carotid arteries are separated by the tumor; accelerated and turbulent flow in the vessels of the tumor). CCA: Common carotid artery; ICA: Internal carotid artery; ECA: External carotid artery; CBPG: Carotid body paraganglioma.

(d) Power Doppler ultrasonography demonstrated in all cases the intratumoral increased vasculature as an abundant flow, characterized as an intense blush pattern, throughout the entire tumor, mainly in small blood vessels. The ECA and ICA were noted to surround the highly vascularized tumor.

Consequently, all our CBPGLs appeared on duplex ultrasound as a hypoechoic, inhomogeneous, well defined and highly vascularized mass arising at the carotid bifurcation between the ICA and ECA, widening the bifurcation without infiltrating it.

MRI/MR-A characteristics

(a) MRI showed a densely enhanced tumor at a widened carotid bifurcation in all patients. On the MR images of three CBPGLs, the lesions were of intermediate intensity on T1-weighted images and slightly hyperintense on T2-weighted images (Figure 2).

MRI indicated the full extent of the tumors and provided information about the absence of infiltration of adjacent structures in all cases, allowing us to assess the entire extent of all CBPGLs.

(b) MR-A confirmed the diagnosis, demonstrating a hypervascularized tumor in the carotid bifurcation. The typical feature was splaying of the carotid bifurcation, with the ECA displaced anteriorly and the ICA and internal jugular vein located posteriorly. All CBPGLs derived their blood supply from the ECA (Figure 3).

Figure 2 – MRI of neck – axial T2-weighted images after Gadolinium: carotid body tumor at left carotid bifurcation. Hypervascular carotid space mass (arrow) splaying the internal and external carotid arteries. Typical “salt and pepper” aspect with prominent vascular flow voids.

Using the preoperative imaging techniques, all neck tumors were classified into group II (3–5 cm) according to their diameter. No lymph node metastasis was found at preoperative imaging in any of the cases.

Surgical treatment

No preoperative embolization of the feeding vessels by superselective carotid artery angiography or fine needle aspiration of the tumor were performed before surgery in any case.

All interventions were realized under general anesthesia. All patients were operated through a longitudinal cervical incision made along the anterior border of the sternocleidomastoid muscle; the internal jugular vein was identified and the common facial vein ligatured. The CCA was dissected to obtain proximal control. Intra-operatively, on exploration of the neck, all seven patients showed CBPGLs group II Shamblin (tumors of medium size, with a latero-lateral diameter between 3 and 5 cm that partially surrounded the carotid vessels or adherent to them). Therefore, we proceeded to a complete subadventitial resection of the tumor in all cases. No lymph node involvement was found intra-operatively in any patient.

Macrosopically, all CBPGLs resected were well-circumscribed, rubbery and reddish brown.

Histological examination

The CBPGL was confirmed in each case on histopathology (Figure 4) and immunohistochemistry, where all the tumor cells tested positive for synaptophysin and chromogranin, negative for cytokeratin. It showed no signs of malignancy in any of the seven tumors. All resected lymph nodes resulted negative for metastasis at histological examination.
Surgical complications and outcomes

There was no operative mortality, and no peri-operative stroke/TIA or severe bradycardia or hypotension were observed. No revision was needed for bleeding (no major bleeding/vascular injury were noted). The operating time was 123±20 minutes. No post-operative complications (respiratory failure, hematoma, infection, or cranial nerve injury) were noted in six cases. The seventh patient presented a transient ipsilateral vague nerve deficit (the tumor was tightly adherent to the neighboring vagus and was successfully resected sparing the nerve, but post-operatively the patient complained of dysphonia). Complete recovery was seen in her case after three months. During the third year follow-up, it was observed that none of our seven patients developed local, regional or distant metastases (recurrence rate was null).

Discussion

CBPGLs, which are hypervascularized tumors of the carotid body [4, 5, 7, 10, 11], constitute the most common form (50%) of head and neck paragangliomas [2, 3, 6–8].

Sometimes, they are multi-center tumors (4%) [2, 7], just like our first case (with an additional glomus tumor of the right prelachrymal sac), being more common in patients with a familial history (7–10% of all CBPGLs) [2, 3, 6–8], like in our first patient. The incidence of bilateral CBPGLs is approximately 5–10% [3, 4, 12], but none of our cases was bilateral. On the other hand, sex prevalence is controversial [13]. In the literature [2, 3, 6–8, 10, 12, 14–16], the mean age of clinical onset in patients with CBPGLs is 40 years, with a high frequency between 30 and 60 years [10]; all our seven cases fell within this age range.

Initially, due to its slow growth, the tumor is generally asymptomatic. In few cases, it is functional (secretes vasoactive substances) [1, 9, 13, 17–20]. We had no clinical symptoms associated with catecholamine production (such as uncontrolled fluctuating hypertension, blushing, and palpitations) in any of the cases.

Then CBPGLs are clinically manifested as palpable painless (non-tender) lateral neck masses (all our seven cases), like vagal paragangliomas [7, 8], located just anterior to the sternocleidomastoid muscle at the level of the hyoid bone [3, 4]. All our seven neck tumors were mobile in the lateral plane, but their mobility was limited vertically, like in other studies [3, 4]. Occasionally, the tumor mass may transmit the carotid pulse: our patients showed unilateral palpable pulsatile neck masses [21]. The presence of vascular hummur near the mass is rare, but it may be a sign of severe carotid artery compression [1, 9] (none of our patients).

In time, the tumor grows and causes external compression, and/or involvement of the surrounding anatomical structures (carotid arteries plus the sympathetic chain, IX–XII cranial nerves, etc.), and manifesting as Horner’s syndrome, dysphagia, etc. [1, 9, 13, 18–23]. None of our cases presented these symptoms before surgery. Jugular paragangliomas present skull base extension, with symptomatic relief [24] (none of our cases).

Other differential diagnosis of CBPGLs is with vagal paragangliomas, thyroid nodules, lymphadenopathy, brachial cysts, carotid artery aneurysms, and salivary gland tumors [24, 25].

Due to the hypervascularization of these tumors, and the proximity to different nervous and vascular structures, fine-needle aspiration biopsy as a preoperative diagnostic tool is not advised as it may cause massive hemorrhage, pseudaneurysm formation and carotid thrombosis [1, 13]. This procedure also presents a risk of dissemination [1, 13]. On the other hand, cytological evaluation cannot differentiate benign from malignant lesions [17, 24, 25]. We did not use this technique in any of our cases.

As direct biopsy is not suitable for the diagnosis of CBPGLs (they are tumors with no clear histological features of malignancy), different imaging modalities are essential in their preoperative diagnosis [1, 9, 13, 18–25].

The use of different types of non-invasive imaging techniques can provide correct preoperative diagnosis in most cases with satisfactory sensitivity and specificity [15, 17, 20, 24–26]. The Joint Vascular Research Group (JVRG), in their meta-analysis study [27], recommend that duplex ultrasound be the primary diagnostic investigation in carotid body tumors. Also used for their preoperative assessment are digital subtraction angiography (DSA), computed tomography (CT), CT angiography (CT-A), MRI and MR-A [20, 27].

Duplex ultrasound helps define vascularity of the tumor (data regarding blood flow in the mass itself), and precise tumor location at carotid bifurcation [1, 22]. Thus, the tumor is represented by a highly vascularized (intralesional abundant blood flow signals) hypoechogenic mass in the area of the carotid bifurcation, which usually causes wide splaying of the bifurcation and separation of the ICA and ECA [1, 2, 7, 22, 28]. All these signs were identified in our seven patients.

Consequently, based on the two main characteristics of the lesion: vascularity and location, duplex ultrasound is helpful in differentiating CBPGLs from other solid, non-hypervascular masses [1, 2, 7, 22, 28–31]. Duplex ultrasound is also suitable for classification of cervical paragangliomas as carotid body, vagal or jugular paraganglioma based on the topographic relation to the carotid arteries and internal jugular vein [1, 22].
of the intrinsic tumor vasculature proved an additional distinguishing criterion upon duplex ultrasound [1, 22]. This technique completely depicts CBPGLs but fails to delineate the high cervical portion of vagal and jugular paragangliomas [1, 22]. Anterior displacement of both carotid arteries and posterior displacement of the internal jugular vein can be found in vagal paragangliomas [1, 22]. The caudal tumor extension of the jugular paragangliomas can be recognized within the expanded lumen of the internal jugular vein [1, 22]. On duplex ultrasound, intratumoral flow is directed cranially in CBPGLs and inferiorly in vagal and jugular paragangliomas. However, there are difficulties with other entities, like ganglion’s metastasis of thyroid or breast cancer [1, 22].

In addition to the diagnosis and differential diagnosis of carotid body tumors, duplex ultrasound can provide information on carotid body tumor size, blood supply of the tumor (feeding arteries) and coexistent carotid artery disease, which is useful in forming a treatment plan and assessing the risk of surgery [1, 22]. On the other hand, Duplex ultrasound has a limited ability to identify complex blood supply to a large carotid body tumor [1], and to measure the tumor exactly [1, 22].

Finally, duplex ultrasound may be useful in screening for familial CBPGLs (like in our first case), and in the follow-up, where it can be a particularly valid first-line tool, thereby reserving second-line techniques such as MRI and CT for selected cases, where it is important to know the accurate dimensions of the lesions [18, 22]. This technique is inexpensive, safe, accurate, readily available, and it is the non-invasive choice for the primary diagnosis of CBPGLs based on its vascularity and location [1, 7, 22, 28–31].

MRI shows characteristic imaging features of CBPGLs that are helpful for their differential diagnosis (“salt and pepper” pattern) [7]. This classic imaging appearance of these lesions on MRI-T2 weighted images, where the “pepper” refers to the low signal flow voids and the “salt” refers to high signal foci of hemorrhage and/or slow flow [9]. This imaging appearance was identified in three of our cases. MRI is better in differentiating of CBPGLs from other paraganglioma [9].

MRI and MR-A with contrast medium administration of the neck are sensitive to assess the presence of tumor at the carotid bifurcation (additional information about the exactly tumor size) (all our cases). They also identify the relationship of the tumor with the adjacent structures (splaying of the carotid bifurcation) [1, 9, 22, 24] (all our cases), and an eventual vascular encasement [13] (any patient). MR-A provides that CBPGL displace the ICA, but do not cause obstruction of this vessel [3] (all our cases). MR-A leads, sometimes, to the identification of an afferent vessel of the tumor (all cases), which could be selective embolized (any patient) [2].

MRI and MR-A are considered as the gold standard imaging techniques for the evaluation of carotid space tumors, as they allow a multiplane approach which is very important in the preoperative study [2, 7, 10, 16, 27, 31, 32]. These techniques are more effective non-invasive imaging modalities compared to with duplex ultrasound, especially for small tumors (diameter <3 cm) [1, 9, 13, 18–23, 31, 32].

Unfortunately, MRI/MR-A do not provide data about the potential for malignancy and postoperative early recurrence because the tumors are too small in terms of the MRI/MR-A’s resolution power [9].

Moreover, both duplex ultrasound and MRI can predict the Shamblin classification, according to several authors [9]. Therefore, both modalities were ordered for all our seven patients.

Digital subtraction angiography (DSA) confirms the diagnosis of CBPGLs demonstrating a hypervascularized tumor in the carotid bifurcation (widening of the carotid bifurcation by a well-defined tumor blush (“lyre sign”), which is classic path gnomonic angiographic finding) [1, 4, 9, 11, 13, 18–23]. The DSA demonstrates accurately the tumor blood supply (it can identify the feeding vessels of the tumor, usually originating from the pharyngeal artery, or CEA). This technique has to be used only in selected cases, to detect the vascular anatomy of large tumors, with identification of an afferent vessel of the CBPGL, followed by preoperative selective embolization of the tumor [13, 19]. For evident reasons, we did not used DSA in any case.

Taking into consideration the practicability and invasiveness of these investigations, as well as the risks and costs involved, and last, but not least, the international literature [7, 8, 17, 25, 30, 31], we propose that the imaging techniques for diagnosis of CBPGLs be performed in the following sequence: (a) duplex ultrasound, (b) MRI, and (c) MR-A.

In consequence, the localization of the tumor within the carotid bifurcation, splaying of the bifurcation, and profuse vascularity are important in the imaging diagnosis of carotid body tumors [1, 9, 13, 18–23].

The selection of treatment depends on the biological activity, size, volume, location, anatomical relationships of the tumor to neighboring structures, such as carotid vessels, craniofacial nerves, the angle of the jaw and the vertebral bodies, as well as the overall fitness of the patient [9].

When the tumor is considered functional, careful clinical evaluation before surgery, measurement of serum catecholamine, treatment with adrenergic blockers, and gentle manipulations during the excision are essential for optimal results [9] (no functional tumor was found in any of our cases).

Given that the natural history of CPPGLs is believed to be unpredictable, immediate surgical removal is recommended. Complete surgical excision remains the golden standard of therapy, as the tumor has a 5% or greater incidence of metastases; radiation therapy and chemotherapy are unsatisfactory [1, 9, 15].

General anesthesia is routine for safe CBPGLs surgery [17], and we used it in all our cases.

The surgical treatment depends on multiple factors, the proper preoperative classification of CBPGLs being imperative for optimal management [1, 9, 13, 18–23, 33].

Based on the relationship to the carotid arteries, Shamblin et al. divided carotid body tumors into three groups [21, 33]. The size of the tumor is positively correlated with the Shamblin classification (the relationship to the carotid arteries), because CBPGLs become more adherent to carotid vessels as they become larger [1, 9, 13,
18–23, 33]. They classified as group I the small tumors, which do not involve the carotid vessels and that can be easily dissected away from the vessels. Group II included CBPGLs of medium size that partially surrounded the vessels or were adherent to them, but could be separated with careful sub adventitial dissection (all our seven patients). Group III consisted of tumors that were large and typically encased the carotid artery, thus requiring partial or complete vessel resection and replacement (ligation of ECA and ICA reconstruction with resection of the tumor) [33].

Luna-Ortiz et al. [33] considered that the Shamblin classification predicts only vascular morbidity without remarking on the neurological morbidity and reflects only the surgeon’s experience in dissecting the tumor. Consequently, they modified the Shamblin grouping as follows: group I – less than 4 cm in size with no surrounding or infiltrating the carotid and excision done without difficulty, group II – more than 4 cm in size with partial surrounding or infiltrating the carotid and excision done with difficulty (all our patients). Group III includes tumors with infiltration to any carotid vessel, more than 4 cm in size, and intimately infiltrating or surrounding the carotid vessels, requiring vascular repair, sacrifice or vessel replacement [21, 23, 27].

Different surgical techniques are used to reduce adverse outcomes and to achieve optimal results as well [1, 9, 13, 18–23, 33]. Careful dissection of the neighboring nerves is essential [9]. Boscarino et al. asserted that the risk of intraoperative cranial nerve injury proportionally increases with the size and extension of tumor (like in our seven patients, which presented the greatest tumor from our seven cases) [13, 19]. For this reason, it is mandatory to assess the preoperative status of neighboring cranial nerves (vagus nerve, hypoglossal nerve, and the superior laryngeal nerve) that are potentially at risk of intraoperative injury [13]. Also, accurate dissection with excision of the tumor should be performed along the subadventitial plane or “white line” suggested by Gordon-Taylor [9, 13, 19] in order to separate the tumor from the surrounding vessels (all our patients).

The surgical procedure must include regional lymph nodes with enlarged size, suspicious morphology, or closely adherent to the tumor [9, 13, 19]. No malignancy was detected in our cases.

Resection of large CBPGLs – group III Shamblin (none of our cases) can be difficult to perform because of their site, size, carotids adherence, local cranial nerve involvement, and hypervascularity, with possible excessive blood loss [15]. A combined therapeutic approach, with eventually preoperative selective embolization followed by surgical resection by an experienced team, offers a safe and effective method for complete excision of such tumors, with a reduced morbidity rate [15, 16, 24, 32]. Embolization is recommended in a few selected cases (size <5 cm, Shamblin’s type III, or significant cranial extension) to decrease the vascularity of the tumor and lower operative blood loss thereby reducing technical difficulty [13, 19]. However, preoperative embolization is still controversial as it may cause ICA thrombosis [1]. No preoperative embolization was performed in our patients.

Histologically, carotid body paraganglioma resemble the normal architecture of the carotid body. The tumors are highly vascular, and between the many capillaries are clusters of cells called zellballen [13, 19] (all our cases).

Concerning postoperative outcomes, surgery has been associated with several complications such as stroke, cranial nerve injury, and bleeding; the risk of postoperative stroke in CBPGLs resection is now less than 5% [4, 13, 19, 27, 33, 34] (no stroke occurred in our cases). However, the incidence of cranial nerve injury remains strikingly high, ranging from 20% to 50%, with larger tumor of Shamblin III variety [4]. Speech and swallowing difficulties could be produced in the immediate postoperative period [9]. In 20% of patients, the neurological deficits are permanent [4]. Postoperatively, we found only one case of transient vague nerve damage. A serious problem during tumor excision is bleeding, which sometimes can be massive. Different authors have recommended standardized ICA shunting during CBPGL excision in order to exclude the vascular supply of ECA, reduce the size of the tumor, and guide the resection in difficult cases [4, 26, 33, 34]. We did not use this technique in any case.

Recurrence after complete resection occurs in approximately 6% of patients [4]; in our study, however, there were no recurrences for three years.

Limitations of our study include: (a) the small number of cases presented (all Shamblin II group), (b) the follow-up period after surgery does not permit long-term conclusions regarding the recurrence of such tumors.

Conclusions

Multidisciplinary management (including radiologists, surgeons, otolaryngologists, anesthesiologists, and neurologists) of patients with CBPGLs is essential. Their early diagnosis (with duplex ultrasound, associated with MRI, and MRA), and complete surgical excision are imperative, like we proceeded in all our seven cases.

Conflict of interests

The authors state that there is no conflict of interests.

References

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