Surgical Treatment of Vagal Paraganglioma with Preservation of the Superior Laryngeal Nerve

Kotsis Thomas and Panagitsa Christoforou*
2nd Department of Surgery, Vascular Division, “Aretaieion” Hospital, National and Kapodistrian University of Athens Medical School, Athens, Greece

Keywords: Vagus nerve; Paraganglia; Paragangliomas; Vagus paragangliomas

Introduction

Parangliomas (PGLs) of the head and neck are quite rare tumors, and although the exact estimation of their incidence is difficult, they represent less than 0.5% of all head and neck tumors. They may arise from the carotid body, the vagal, tympanic, jugular paraganglia [1]. Less than 5% of head and neck PGLs originate from the vagus nerve [2]. The cranial nerve X or vagus nerve is the dominant nerve of the parasympathetic division of the autonomic nervous system; the vagus paraganglioma (VPGL) is a primary example of an endocrine tumor associated with the vagus nerve [3].

We present herein a case of a woman with a mass at the left side of the neck, which turned to be a vagal paraganglioma; the mass was surgically excised. Furthermore, the clinical findings, imaging investigations, differential diagnosis and surgical treatment steps are presented; we highlight the importance of early diagnosis and surgical treatment.

Case Report

A 50-year-old woman, with a free past medical history, observed a gradual increase of asymptomatic mass in the left cervical. During the clinical check a solid adherent, compact, painless mass on the inside of the left anterior sternocleidomastoid was found with minimal lateral mobility. The mass was close to the carotid pulse with limited vertical movement. She had no family history of paragangliomas or other neuroendocrine tumors, or any relevant past medical history. Neurologic examination was unremarkable. The initial thought was that of a branchial cleft cyst. The patient underwent a neck ultrasound (U/S) study which revealed a compact, oval mass of 3 x 1 cm, well-defined hypoechoic, solid lesion at the left carotid triangle, with rich vascularity in the power Doppler resembling carotid body paraganglioma (CBPGL). It was followed by magnetic resonance angiography (MRA) (Figure 1), where a mass was found to repel the cervical vessels. A digital subtraction angiography (DSA) followed; a tumor was pressing the internal and external carotid artery just over the carotid bifurcation, demonstrating a vascular “blush” and reversing the carotid division to the middle line (Figure 2).

Figure 1: Magnetic Resonance Angiography (MRA), where a mass was found to repel the cervical vessels (arrow).
The patient was electively admitted in our hospital and underwent a surgical excision of the tumor under general intratracheal anesthesia; the mass and the carotid bifurcation were covered by an arachnoid web-like structure and rich venous web (Figure 3). There was arterial supply from the external carotid artery crossing the carotid bifurcation in the front plane (Figure 4); this artery supplying the tumor was ligated. The tumor was removed, with the usual left lateral cervical incision until the base of the skull. The tumor was removed en bloc with the vagus nerve. Any attempt to separate the tumor from the nerve was fruitless, so we decided to remove it with the part of the nerve to which was adhered; the cranial growth of the tumor allowed the superior laryngeal nerve to be maintained; the two ends of the vagus nerve were ligated (Figures 3-8). The patient experienced a hoarseness, directly postoperative, which occurred for about one month; further investigation with neck ultrasound and brain MRI was performed and was reported as normal. The histopathology showed a 3 cm vagal paraganglioma. She had an uneventful post-operative recovery and was subsequently discharged on the 3rd post-operative day (POD); the regular follow-up check for 7 years after surgery did not indicate any radiological or functional abnormality and showed no evidence of recurrence.

Discussion

Paragangliomas (PGLs) of the head and neck are quite rare tumors, and although the exact estimation of their incidence is difficult, they are regarded to represent less than 0.5% of all head and neck tumors. Overall, there is a 3:1 female prevalence and the two-thirds of the cases are diagnosed between the ages of 40 and 60 [4].

Paragangliomas may arise from the carotid body, vagal, tympanic and jugular paraganglia; they are associated with the parasympathetic nervous system and are typically non-secretory. In contrast, PGLs of thorax and abdomen are secretory and this is the big difference from the PGLs of the head and neck [3,5].

Less than 5% of head and neck PGLs originate from the vagus nerve [2]. The cranial nerve X or vagus nerve is the dominant nerve of the parasympathetic division of the autonomic nervous system. VPGLs are a prime and distinctive example of an endocrine tumor associated with the vagus nerve.

Embryologically, preganglionic motor, somatic sensory neurons and supporting cells of the cranial ganglia, originate from neural crest cells. The vagus nerve descends vertically through the neck within the carotid sheath and within the mediastinum following different paths on the left and right sides. The cell bodies of fibers, which transmit the visceral stimuli, and regulate “rest and digestive” functions of parasympathetic system, are located in the inferior ganglion of the
vagus nerve; it constitutes the pathway of communication between the central nervous system and the viscera transmitting more than 80% of sense [6].

Vagal PGLs may develop anywhere along the course of the vagus nerve, but usually arise from the inferior ganglion of the vagus nerve or the plexiform ganglion lying more cranially and more medially than the carotid body [7].

In the case we present here the tumor had a lateral route, protruded to the carotid bifurcation pressing forward and medially the carotid bifurcation provoking a clear deviation of the internal carotid artery (Figures 2-5).

Vagal PGLs are more commonly found in women [3]. Vagal PGLs are presented as a painless, rubbery compressible, slow growing mass in the upper neck, with limited vertical mobility and free lateral mobility, in the 60% of the cases. The tumor mass may transmit the carotid pulse and if is enlarges around the carotid vessels and X-XII cranial nerves, symptoms, like odynophagia, dysphagia and voice’s hoarseness may occur. As the mass is growing, syncope may be seen due to compression of carotid sinus or internal carotid artery. Another symptom is Horner’s syndrome, which is a rare disorder characterized by a constricted pupil, drooping of the upper eyelid and absence of sweating on the face and is an aftereffect of the compression or the invasion of cervical sympathetic chain. The tumor may be pulsatile and it can express a constellation of symptoms. Vagus is the most usual nerve that paralyzes among the cranial nerve palsies, during operation, and is presented in 30% of the patients [1].

The patient we operated had a mild clinical picture with palpable mass on the left side of the neck, without any other significant symptoms. The patient experienced a hoarseness, directly postoperative, which occurred after one month.

The percentage of 65% of PGLs is sporadic and the remnant is inherited, even if this expression of the mutations in the succinate dehydrogenase subunit D (SDHD) gene is still not fully understood. Young people are the main category where familial PGLs syndromes occur [5,6].

According to the authors [8-11], patients who have more than one of the following characteristics should subsist in genetic testing: age over 50 years, family history, extra-adrenal tumors, multiple or/and metastatic tumors, elevated dopamine methoxytyramine. Immunohistochemistry may also be used for identification of mutations on resected tumors [11].

In the case we described no genetic test was performed due to negative medical history and no family history of paragangliomas or other neuroendocrine tumors.
The paraclinical investigation for VPGLs includes ultrasound, computed tomography (CT) and CT angiography (CTA), magnetic resonance imaging (MRI) and MR angiography (MRA) and conventional arteriography, while functional molecular imaging examination includes somatostatin receptor scintigraphy (SRS), [18F]-FDG PET, [18F]-FDOPA PET, [68Ga]-labelled somatostatin agonist PET.

Computed tomography angiography (CTA) and magnetic resonance angiography (MRA) provide valuable information to determine the diagnosis and preoperative planning, because the lesions strongly reflect their rich vascular nature with contrast. The tumor has a "salt and pepper" appearance using the classic MRI flow voids, while 123I-metaiodobenzylguanidine (123I-MIBG) scintigraphy can be used to distinguish metastatic or convert VPGLs [4].

Despite the progress of MR and CT technology, digital subtraction angiography (DSA) remains the gold standard in estimating tumor vascularity or identifies multiple vascular tumors in the head and neck, and is carried out near the time of planning surgery to allow embolization, if required [12].

Conventional arteriography is performed frequently for the very large VPGLs of the vagus nerve or skull base, given the fact that these tumors have a propensity to skull base or even intracranial extension through the jugular foramen [7]. According to the Netterville-Glasscock classification, which estimates the expansion of the lesion in relation to the distance from the skull base, three groups are listed: A: distant, B: in contact, C: the tumor enters the jugular foramen (Table 1) [13].

Positron emission tomography (PET) should be performed in order to detect multiple tumors or associated thoraco-abdominal tumor; 68Ga-labelled somatostatin agonist PET is also the preferred examination, regardless of the genetic background [7].

In general, when in a neck CT or MRA, a hyper vascular mass in relation to the major cervical vessels is recognized, provoking an anterior/medial displacement of the carotid bifurcation and a posterior displacement of the internal jugular vein, the diagnosis of VPGL must be suspected and a surgical treatment has to accordingly be planned [14].

Differential diagnosis includes neuroma, jugular meningioma, schwannoma, meningioma, carotid paraganglioma, jugulotimpanic Paraganglioma [7, 14].
The early excision has been constituted because of the unknown malignant potential of VPGLs and hypothesis for progressive, local advancement over time; even in the benign setting [15].

Surgical excision is the classical and the only therapeutic option of treatment for the most VPGLs; symptomatic tumors and malignant disease. The difficulty of dissection of the cranial nerves (IX, X, XI, XII) is variable, but the nerves can be difficult to detect among the tumor capsule. Unfortunately, the sacrifice of the vagus nerve is often needed, causing speech, swallow, and sensory deficits [16].

Cranial nerve preservation is unlikely in the presence of an advanced tumor.

The functional complications of VPGLs depend on tumor size; tumors less than 2 cm in diameter can be removed with functional damage limited to the vagus nerve. In the case of multiple nerve lesions the postoperative reparation is complicated, while retrieval of speech and swallowing functions can be elongate, whereas speech therapy can be an important tool for re-speaking as before. Immediate postoperative disorders of swallow like multiple paralysis of X, XII, IX, and superior laryngeal nerve are occurring in a high percentage of 60 to 80%, and are persistent in 50% of cases, requiring feeding gastrostomy. Patients with severe swallowing disorders redo surgery, like thyroplasty, vocal cord injection, gastrostomy or even tracheotomy and airway protection is necessary in order to protect the airways and reduce mortality. Other complications include respiratory complications, severe aspiration pneumonia, cardiac arrhythmias, sinus tachycardia and/or positional blood pressure instability with disabling orthostatic hypotension [16].

The above functional disorders may occur after bilateral laryngeal nerve palsy during excision of bilateral VPGLs, so the need for cranial nerve (IX, X, XII) examination, before treating the contralateral side, is strictly appropriate [17].

Similar, when a carotid bulb tumor and a vagal tumor coexist, but on different sides, the best option is the carotid bulb tumor to be resected firstly, so minimizing the risk of bilateral vagus nerve palsy.

In the case we present here, we attempted to separate the mass from the nerve but it was clear that this was impossible. Although it is reported that in many cases this is possible, in our case this was not feasible; however perhaps due to the preservation of the vagus stem (Figure 7) containing the superior laryngeal nerve origin, no further clinical neurological or vocal problems were detected.

Vagal PGLs are frequently malignant, without preoperative criteria predictive of malignancy to be identified. The frequency of malignancy is difficult to estimate, as the only recognized criteria are the attendance of cervical lymph node metastases and/or distant metastases. Malignant VPGLs disease is determined exclusively by the presence of metastases invading non neuro endocrine tissue.

Surgical resection followed by radiation therapy is the most commonly treatment described for malignant VPGLs, even if their rare occurrence is not clear. The survival rate is approximately 80% for five years, if lymph node metastases are located and 12% with distant metastases.

Conventional, nonsurgical treatment for malignant, nonresectable VPGLs includes radionuclide therapy, alkylating agents or tyrosine kinase inhibitors [15].

More recent methods of radiotherapy, which allow decreased irradiation of healthy tissues, are the gamma knife with single fraction, while conventional stereotactic radiosurgery is based on fractionation in 25 to 30 sessions and the cyber knife, which is a new modality of whole-body stereotactic radiotherapy, allowing the treatment of extra cranial sites, recording 0% of complications [13].

Conclusion

The best policy concerning vagal paragangliomas is a high suspicion index in order to excise the tumor in surgically allowable dimensions. Any effort is aimed at eliminating surgical damage in the cranial nerves, which however occurs in a percentage of 10-30% [18]. Therefore, early detection by modern imaging methods and prompt surgical resection will decrease surgical complications.

References