Case report

Vagal paragangliomas: two case reports

Paragangliomi vagali: descrizione di due casi

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SUMMARY

Two uncommon cases of paragangliomas arising from the vagus nerve are described. The first patient underwent surgery for suspected carotid body tumour. In the second patient, computed tomography scan and digital angiography allowed a correct pre-operative diagnosis to be made. These cases confirm the prevalence of vagal paragangliomas in female sex and middle age, and the possibility of multiple similar tumours in the same patient. Histological benign features, absence of neurological symptoms, of local invasion or intracranial extension confirm the frequent benign behaviour of these neoplasms. Lack of catecholamine secretion confirms the low incidence of functioning tumours. Contrast computed tomography and digital angiography still remain the gold standard reliable instruments for diagnosis despite the success of magnetic resonance imaging, magnetic resonance angiography and octreotide scintigraphy to detect head and neck paragangliomas. A transcervical approach, without mandibulotomy, is suitable too for large tumours but complete removal, with sparing of involved segments of the vagus nerve, is rarely possible. Post-operative neurological morbidity is still an unsolved issue and, therefore, rehabilitation of deglutition and phonation is an integral part of management.

KEY WORDS: Head and neck tumours • Glomus vagal tumour • Vagal paraganglioma • Surgical treatment

Introduction

Vagal paragangliomas are uncommon tumours of the parapharyngeal space that present clinical and pathological characteristics which differentiate them from similar head and neck tumours. Epidemiology, tumour biology and radiologic appearances are well known but some controversies exist in diagnosis and treatment modalities. Pre-operative acknowledgement of the tumour and post-operative neurological deficits prevention are issues still unresolved. Adjunctive procedures to facilitate the rehabilitation of the patients do not seem to reduce post-operative neurological troubles and are not a standardized cure.

In this paper, two cases of benign vagal paragangliomas, situated in the jugulo-digastric area without intracranial extension and neurological signs are described; these were studied by contrast-enhanced computed tomography (CT) and carotid vessels angiography and subsequently treated with surgical removal of the tumour together with the segment of the vagus nerve involved. Clinical presentation, radiological modalities for preoperative diagnosis, surgical treatment and postsurgical morbidity are herewith compared with previously reported data.

Case 1

A 60-year-old female was admitted on February 14th, 1996 because of swelling in the left upper region of the neck. The patient had undergone surgery for resection of a mediastinic paraganglioma in 1988 and of a right carotid paraganglioma in 1995. Excision of the contralateral mass was suggested too but the patient did not accept the procedure at that time. Upon examination, the swelling measured 6 x 4 cm and appeared lobular, firm and fixed to the underlying structures in
the left jugulo-digastric area. The patient presented a scarred incision along the anterior margin of the right sternocleidomastoid muscle and another scar on the anterior chest wall for previous midline sternotomy. Oropharyngolaryngoscopy revealed no medial displacement of peritonsillar structures and no vocal cord paralysis. A previous contrast-enhanced CT scan of the neck, performed in 1995, before surgical removal of the right carotid paraganglioma, revealed lesions on both sides, at level of carotid bifurcation, with high contrast enhancement (Fig. 1). On the left side, a well-defined hyperdense mass (6.4 x 4 cm), which extended cephalad not up to the skull base and medially to the parapharyngeal space, produced asymmetry of the aero-pharyngeal space and bulging of the pharyngeal wall into the pharyngeal lumen. A carotid angiography performed before excision of the right carotid paraganglioma showed, on the left side, a highly vascular mass on the outside of carotid bifurcation (Fig. 2a) with arterial supply from left external carotid artery. On the right side, angiography showed evidence of the removed vascular lesion inside carotid bifurcation (Fig. 2b). A new CT scan was not performed because swelling did not appear increased in the last months and the patient continued to be asymptomatic.

The upper region of the neck was explored through an oblique incision along the anterior border of the sternocleidomastoid muscle extending to the preauricular region. A vascular brownish-red mass was found between the carotid arteries antero-laterally and the internal jugular vein posterior-medially not occupying the carotid bifurcation angle. The tumour, probably originating from the inferior ganglion of the vagus nerve, compressed and dislocated, but did not infiltrate the jugular vein and the hypoglossal nerve from which it was separated, without vascular or neurological lesions. Even though the mass extended cephalad in the neck and medially to the parapharyngeal space, it was possible to approach it by means of a trans-cervical incision with diagastric and stiloid tendon resection and stiloid bone rupture without other manoeuvres. Since the tumour encapsulated the vagus nerve, thus not allowing separation, it was completely excised together with the involved part of the nerve. No further procedures to facilitate post-operative rehabilitation of the patient were performed. Histopathology of the resected specimen was consistent with paraganglioma of vagal origin.

After the operation, the patient suffered from mild paralysis of the left facial nerve, hoarseness and nasal regurgitation. Fibre-optic laryngoscopy revealed paralysis in abduction of the left vocal cord and the patient underwent deglutition and phonation rehabilitation. At 6-month follow-up, the patient was found to have less evident signs of left facial nerve paralysis, and no longer presented aspiration, but still suffered from persistent hoarseness of the voice. Today, 8 years after surgery, the patient is well, not presenting any recurrence or metastases and, moreover, has an almost normal voice.

Case 2

A 46-year-old female was admitted on July 5th 2004 with a 12-month history of painless swelling in the left upper region of the neck. Patient examination revealed a small, painless, firm, swelling 3 x 2 cm in size, in the left jugulo-digastric area. Contrast enhanced CT scan of the neck revealed a hypervascular lesion 31.4 x 15.6 mm in size, which compressed the internal jugular vein and displaced anteriorly the carotid vessels (Fig. 3a, b). A carotid angiography confirmed the presence of a highly vascular mass lying outside and above the carotid bifurcation (Fig. 4a), with an arterial supply from the external carotid artery by means of a hypertrophic ascending pharyngeal branch (Fig. 4b). Magnetic Resonance Imaging (MRI) was not performed for the presence of ferromagnetic artifacts due to a not removable dental prosthesis.

During surgery, performed by means of a trans-cervical approach, a 3 x 2 cm highly vascular lesion was found displacing forward the internal carotid artery and backwards the jugular vein without occupying the carotid bifurcation angle. The lesion, found to arise from the left vagus nerve, was excised together with part of the nerve involved (Fig. 5a, b). No additional procedures to improve rehabilitation were carried out. The histopathology of the resected specimen
revealed polygonal neoplastic cells with round/oval-shaped nuclei, infiltrating capillaries and extensive necrosis, thus confirming the diagnosis of vagal paraganglioma. Post-operative morbidity revealed hoarse voice and some difficulty in swallowing, from which the patient promptly recovered within a week. Subsequent laryngoscopies demonstrated persistence of the post-operative left vocal fold paralysis.

Discussion

Vagal paragangliomas are unusual tumours (approximately 200 cases having been reported) arising from nests of paraganglionic tissue within the perineurium at various sites of the vagal nerve, but generally in the region of the jugular and nodose ganglia. They account for only 3% of all head and neck paragangliomas and are the third in prevalence after carotid and jugular localization. As opposed to carotid body tumours, vagal paragangliomas are located more cephalad in the neck, between the jugular vein and the internal carotid artery, sometimes extending to the base of the skull through the jugular foramen or posteriorly to the mastoid tip. Intracranial extension, which is the main cause of death, occurs in 22% of the cases. Oropharyngeal involvement, due to bulging of the pharyngeal wall into the pharyngeal lumen and medial displacement of the tonsil, may occasionally be observed.

Vagal paragangliomas which occur more frequently in female (2.7:1 ratio) and middle age patients have a slow growth rate. Typically, they present with an asymptomatic neck mass behind the angle of the mandible, with no other symptoms. In less than 50% of cases, or even less frequently, they present with cranial neuropathies such as paralysis of the 10th, 9th, 11th, 12th cranial nerves, which manifest as hoarseness of the voice, dysphagia and shoulder drop, nasal reflux of fluids and aspiration, hemiatrophy of the tongue and unilateral cord paralysis. Vagal paragangliomas may be sporadic or familial, with an autosomal dominant pattern with variable penetrance. The incidence of secreting tumours (paroxysmal hypertension, headache, palpitation and sweating) is estimated to be less than 1-3% of all head and neck paragangliomas; 19% manifest malignant behaviour destroying adjacent structures and 10-20% metastasize to regional lymph nodes or to lungs and/or brain cranial structures.

Histologic features are similar to those of other paragangliomas: nests of epithelioid or polygonal cells, homogeneous or with cellular atypias, mitosis and capsular invasion, surrounded by reticular fibers and many blood vessels in an arrangement called “Zellballen” pattern. Since there is no correspondence between histologic characteristics and clinical behaviour, malignancy is not a histological, but a clinical, diagnosis, made when local invasion or metastases are present.

Fig. 3a, b. Contrast-enhanced CT scan of neck showing left mass 31.4 x 15.6 mm compressing internal jugular vein and displacing anteriorly external and internal carotid arteries.

Fig. 4. Digital angiography showing a tumour blush lying outside and above the carotid bifurcation (a) with feeders from external carotid artery particularly from a hypertrofied ascending pharyngeal artery (b).

Fig. 5. Operative view showing vagal tumour involving vagus nerve (a) and removal with segment of vagus nerve involved (b).
Pre-operative diagnosis of vagal paraganglioma, even though difficult, can be made combining clinical findings and radiographic studies. High definition imaging techniques (contrast computed tomography and conventional angiography) reveal the tumour as an intensely vascular enhanced mass, which displaces the internal and external carotid arteries anteriorly and the internal jugular vein posteriorly. Apart from vascular displacement, carotid angiography may demonstrate tumour arterial supply by feeders from the external carotid artery and particularly from the ascending pharyngeal artery. However, at present, MRI is considered the most important imaging modality, especially for lesions involving the skull base. Octreotide scintigraphy can identify occult, distant lesions and small recurrences and can also measure tumour secretory function, in a few functioning tumours. Diagnosis of secreting paraganglioma involves the use of a screening test for serum catecholamines and a urine test for catecholamine metabolites.

The treatment of choice for these tumours is surgery which generally requires sacrificing the vagus nerve and is associated with other nervous lesions. Therefore vagal, hypoglossal and glossopharyngeal paralysis are common postoperative complications. Therapeutic embolization reduces the size of the tumour and limits intra-operative blood loss but may rarely be considered as definitive treatment. Radiation therapy is an alternative to surgery and may be effective in arresting the growth, but it can lead to neurologic sequelae and rarely can destroy the tumour. Irradiation is generally used in elderly patients as well as in large and unrespectable tumours. Monolateral radiotherapy can be indicated after controlateral surgery in order to avoid bilateral vagus nerve damage.

These two case reports are examples of vagal paragangliomas of the jugulo-digastric area, probably arising from or near the inferior ganglion of the vagus nerve, without invasion of the skull base and lacking in neurological and malignant signs. The origin of these tumours is presumed, since only tumours arising from or near the inferior ganglion are situated behind the angle of the mandible between the internal carotid artery and the jugular vein. Painless neck swelling, with no other symptoms, do not concur with published statistics which reveal a 50% cranial nerve paralysis at patient presentation but with those statistics which reveal a lower frequency. Our male/female ratio of 0:2, an average age of 53 years and an average size of the tumour of 4.9 x 3.5 cm on presentation, correlate well with the corresponding figures reported in the literature.

The history of our first patient, with two previously resected paragangliomas, confirms the possibility of multiple localization which is considered to occur in 10-20% of non-familial paragangliomas and in 33-50% of familial paragangliomas. As far as this aspect is concerned, we have found no other cases, in the literature, of synchronous or metachronous mediastinic, carotid and glomus vagal paragangliomas, but only carotid-vagal, carotid-jugular, vagal-vagal or carotid-vagal and glomus tympanicum combinations. Glomus jugulare and glomus tympanicum, with carotid body tumour, are the most common combinations. Despite multicentricity, this patient did not have a familial history of paragangliomas. The second case was also an example of sporadic non-familial tumour, which occurs in approximately 8% of these patients.

Despite pre-operative radiographic studies (contrast-enhanced CT and digital angiography), in our first case, the vagal origin of the tumour was a diagnostic surprise; the exploration was performed based on clinical suspicion of a carotid body tumour. Probably, the histology of the right neoplasm, previously removed, and the erroneous idea that bilateral paragangliomas arise from the same structure on both sides caused us to make this mistake. In this patient, angiographic signs were not correctly evaluated because the anterior displacement of the internal carotid artery by the intensely enhanced mass and the not enlarged bifurcation angle should have been evident. Differences from the right carotid body tumour appearance with enlarged carotid bifurcation should also have been evident. A similar bilateral cervical paraganglioma was described in a recent report in this case the tumour arose from the vagus nerve on the right side and from the carotid body on the left side. This patient, however, did not present a third localization in the mediastinic region. In our second case, contrast-enhanced CT and angiogram clearly documented the vascular mass more cephalad in the neck and more lateral with respect to the carotid bifurcation than usually occurs in carotid body tumours, compressing the internal jugular vein and displacing forward the internal carotid artery, with a non-enlarged carotid bifurcation angle, thus correct diagnosis was made pre-operatively.

In our two patients, surgical treatment was performed by means of a simple trans-cervical approach because the first neoplasm, despite its large dimensions, was not extended very cephalad and did not invade the skull base and the second tumour, smaller in size, was strictly limited to the jugulo-digastric area. At surgery, in the first case only a minimal disruption of the normal anatomic structures was performed limited to digastric and styloid muscle resection and styloid apophysis rapture. Neither midline mandibulotomy, nor temporal bone destruction or posterior fossa craniotomy was necessary in both cases presenting massive invasion of the parapharyngeal space or with intracranial extension of the neoplasia. In these between the internal carotid artery and the jugular vein. Painless neck swelling, with no other symptoms, do not concur with published statistics which reveal a 50% cranial nerve paralysis at patient presentation but with those statistics which reveal a lower frequency. Our male/female ratio of 0:2, an average age of 53 years and an average size of the tumour of 4.9 x 3.5 cm on presentation, correlate well with the corresponding figures reported in the literature.

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Post-operative ipsilateral vocal cord paralysis produced, in both patients, significant post-operative problems such as preservation of anatomical integrity is not synonymous with function preservation. Neither midline mandibulotomy, nor temporal bone destruction or posterior fossa craniotomy was necessary in both cases presenting massive invasion of the parapharyngeal space or with intracranial extension of the neoplasia. In these between the internal carotid artery and the jugular vein. Painless neck swelling, with no other symptoms, do not concur with published statistics which reveal a 50% cranial nerve paralysis at patient presentation but with those statistics which reveal a lower frequency. Our male/female ratio of 0:2, an average age of 53 years and an average size of the tumour of 4.9 x 3.5 cm on presentation, correlate well with the corresponding figures reported in the literature.

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are well known causes of post-operative aspiration. In our two patients, continuous post-operative rehabilitation resolved aspiration after a few days and hoarseness and dysphagia after some a few months, whereas the vocal cord in abduction paralysis has, so far, shown no improvement. In conclusion, this report confirms the prevalence of vagal paragangliomas in the female sex and middle age, the possibility of multiple tumours in the same patient, their frequent benign behavior and the low incidence of functioning tumours. Contrast-CT and conventional angiography still remain reliable instruments in the diagnosis of these lesions, especially in patients having contraindications to MR. Digital arteriography is still of great value in revealing the angioarchitecture of the tumour and in detecting multiple tumours which have escaped MRI. Radiological findings of a more cranial and lateral lesion, with respect to the carotid bifurcation, and the non-enlargement of the bifurcation angle, can differentiate vagal tumour from more frequent carotid paraganglioma. Complete tumour removal without sparing of the vagus nerve is rarely possible and post-operative rehabilitation of cranial nerve deficits must be considered as an integral part of management.

References


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