Hemiatrophy of the Tongue Caused by an Extensive Vagus Nerve Schwannoma Masquerading as a Carotid Chemodectoma

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Abstract

Background Carotid chemodectomas or carotid body tumors are rare neoplasms, generally benign and frequently asymptomatic, which represent 0.6 % of all head and neck tumours. Schwannomas or neurilemmomas are benign, slow growing tumours that derive from the sheath of peripheral and cranial nerves. Of all neurilemmomas, 25–45 % originate from the head and neck region, with the vagus nerve being one the most encountered sites of occurrence.

Case Report We describe a schwannoma originating from the vagus nerve mimicking a carotid body tumour and compromising the function of the hypoglossal nerve.

Discussion Patients with lateral neck masses near the bifurcation of the carotid artery are often referred to the maxillofacial surgeon. The differential diagnosis of these lesions includes a variety of tumours among which carotid chemodectomas and vagus nerve schwannomas have a significant incidence. Both lesions may appear as completely asymptomatic, thus it is very difficult to identify the origin of the tumour based on physical examination. In making a differential diagnosis of the cervical tumours imaging studies play a key role.

Purpose A diagnostic algorithm based on contemporary imaging modalities, which can be useful in order to distinguish between these two pathological entities is presented.

Keywords Vagus nerve schwannoma · Carotid chemodectoma · Hemiatrophy of the tongue

Introduction

Carotid chemodectomas or carotid body tumors are rare neoplasms, generally benign, slow-growing and frequently asymptomatic, which represent 0.6 % of all head and neck tumours [1]. They usually present in the neck, anterior to the sternocleidomastoid muscle at the level of the hyoid bone, near the carotid bifurcation and can be pulsatile due to their juxtaposition with the carotid artery. These tumours are functionally active only in 1–3 % of the cases and malignant transformation may occur in 3–12.5 % of all lesions [2]. Metastases are found in less than 5 % and multiple tumors in 5 % of the cases, respectively. The demonstration of neurological deficit caused by vagal or hypoglossal nerve involvement is a rare phenomenon and the occurrence of Horner syndrome is even more uncommon [3].

Schwannomas or neurilemmomas are benign tumours that derive from the sheath of peripheral and cranial nerves. Of all neurilemmomas 25–45 % originate from the head and neck region [4], with the vagus nerve and sympathetic chain being the most encountered sites of occurrence [5]. These neoplasms when developed in the neck usually present as asymptomatic, solitary, painless masses with a slow pattern of growth. Malignant transformation of schwannomas, as well as the spread of the tumour to other cranial nerves is considered to be a rarity [6]. We present a case of vagal neurilemmoma mimicking a carotid body tumour, affecting also the hypoglossal nerve and discuss a differential diagnosis algorithm in order to facilitate the diagnostic process of these lesions.

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Report of the Case

A 34-year old male presented with a well-defined swelling of his right neck, increasing hoarseness, and rightward tongue deviation on protrusion present for 15 months. Physical examination revealed a right lateral neck mass extending from the superior aspect of the sternocleidomastoid muscle to the angle of the mandible with concomitant right tongue hemiatrophy and impairment of lingual mobility (Fig. 1). The lesion was elastic, non-tender and mobile horizontally but not cephalocaudally. The function of the cervical sympathetic chain was intact. No palpable cervical lymph nodes were present. The patient's medical history was unremarkable.

Computed tomography (CT) scan with contrast showed a well-defined $4 \times 3 \times 6$ cm mass, located between the internal carotid artery and the external carotid artery splaying the bifurcation and compressing the internal jugular vein (Fig. 2). Intravenous injection of contrast medium also demonstrated feeding vessels of the tumour originating from the external carotid artery. Magnetic resonance imaging (MRI) scan revealed a contrast enhancing mass within the right carotid canal extending from the angle of the mandible to the carotid bifurcation (Fig. 3). Fine needle aspiration cytology (FNAC) was not performed in order to avoid the risk of penetrating a vascular tumour.

At the time of the operation, it was determined that the tumour was located posterolateral to the bifurcation, appearing to arise from the vagus nerve (Fig. 4). The lesion separated the carotid arteries and the internal jugular vein which was also compressed. The right hypoglossal nerve was severely compressed. The mass was encapsulated and could be resected enbloc since the vagus nerve was not coursing through the tumour but was passing over the tumour capsule. Therefore, the nerve was preserved.

Fig. 1 Preoperative presentation of patient showing deviation of tongue and right hemiatrophy

Fig. 2 Axial CT of the neck showing the mass which caused splaying of carotid bifurcation

Fig. 3 Magnetic resonance imaging (MRI) scan reveals a contrast enhancing mass within the right carotid canal extending from the angle of the mandible to the carotid bifurcation

Fig. 4 Intraoperative photo showing the exact position of the tumor (has just been removed) on the carotid bifurcation

Fig. 5 S-100 stain (strongly and diffusely +) especially in Antony A areas. Highly specific for schwannoma

Fig. 6 Vimentin stain (+) non specific for schwannoma

Histopathologic examination revealed a benign Antony A and Antony B neurilemmoma with positive staining for S-100 and vimentin (Figs. 5, 6). Patient's postoperative course was uneventful and 6 months later the mobility of the tongue was significantly improved. His voice was back to normal, with no clinical signs of aspiration to be present.

Discussion

The parapharyngeal space is divided by the styloid process into an anterior and a posterior compartment. The posterior compartment, which is surgically more interesting, contains the internal and external carotid arteries, the cervical sympathetic chain, the internal jugular vein, the glossopharyngeal, vagus, spinal accessory and hypoglossal nerves and the tail of the parotid. Therefore, the differential diagnosis of a lateral neck mass includes: salivary gland tumours, vascular tumours and malformations, neurogenic tumours, lymphatic masses, lipomas, thyroid masses and branchial cleft cysts.

Vagal schwannoma is typically characterized by dysphagia and hoarseness. Occasionally, a paroxysmal cough may be produced on palpation of the mass. Carotid chemodectomas usually present as a painless mass in the lateral neck which can be pulsatile. Usually the mass can be moved laterally but not cephalocaudally since it adheres to the carotid. Also sounds and murmurs can be heard through auscultation. Tumour's progressive growth may cause symptoms like dysphagia, odinophagia and hoarseness because of the anatomical proximity of the vagus and hypoglossal nerves with the carotid vessels. However, both lesions may appear as completely asymptomatic, thus it is very difficult to identify the origin of the tumour based on physical examination. In making a differential diagnosis of the cervical tumours imaging studies play a key role. The role of the FNAC in this process is less valuable and most would give an inconclusive result, therefore it is not recommended by some authors [7].

When a schwannoma of the vagus nerve is enlarged, the common carotid artery is displaced medially and the internal jugular vein laterally [8]. On the contrary, the anatomic origin of a carotid body tumour that increases in size, indicates that a splaying of the carotid artery bifurcation will occur, separating the internal from the external branch. Additionally, schwannomas are generally avascular lesions whereas chemodectomas tend to be vascular tumours [9]. On computed tomography, splaying of the carotid bifurcation caused by a hypervascular mass almost always indicates a carotid body tumour. Separation of the internal jugular vein from the internal carotid artery, without splaying of the carotid bifurcation usually indicates a vagal schwannoma [10]. In our case, these two elements

of carotid bifurcation splaying and hypervascularity of the mass led to the working diagnosis of a carotid body tumour rather than a schwannoma. This hypothesis was further supported by the fact that there are clear suggestions in the literature that a vagal nerve schwannoma could never separate the internal and external carotid arteries [7, 10].

Magnetic resonance imaging is considered the modality of choice for the evaluation of parapharyngeal space tumours. Miller et al. [11] showed that MRI can delineate the relationship of a parapharyngeal mass to the parotid gland and major vessels of the neck, as well as significant lesion characteristics such as vascularity or solidness with an accuracy of 95 %. On MRI, neurilemmomas are well-defined homogenous masses exhibiting high signal intensity on T₂ weighted images and a low signal intensity on T₁ weighted images. In contrast with chemodectomas, schwannomas do not demonstrate vascular flow voids on MRI [12]. In the presenting case, intravenous injection of contrast medium revealed feeding vessels of the tumour originating from the external carotid artery, enhancing the impression that the mass was a carotid body tumour. Chronic and significant compression of the hypoglossal nerve may cause hemiatrophy of the tongue but this has been rarely reported [13, 14]. However, the etiology of hemiatrophy in both presented cases was a hypoglossal nerve schwannoma. To our knowledge the only case of a vagal schwannoma causing hypoglossal nerve compromise and concomitant hemiatrophy of the tongue was described by Schupp et al. [9]. Our patient initially presented with a rightward tongue deviation on protrusion, hemiatrophy and impairment of lingual mobility, symptoms that were consistent with chronic compression of the twelfth cranial nerve. After the tumour was removed, the hypoglossal nerve was decompressed which resulted in the improvement of lingual mobility 6 months postoperatively.

There is universal agreement that encapsulated schwannomas are completely benign and very rarely undergo malignant transformation [6, 9]. Hence, complete surgical excision of the tumour, which remains the treatment of choice, should preserve the integrity of the nerve when possible. These tumours arise outside the involved fascicle, tend to compress the nerve fibers and can usually be dissected free [12, 15]. Enucleation of the tumour from the nerve is desirable in order to preserve its function. However, if complete tumour removal requires sacrifice of a portion of the nerve of origin, an end to end anastomosis or an interposition nerve grafting should be performed. In any case, patients should be informed for dysfunction of the involved nerve during preoperative consent.

Patients with lateral neck masses near the bifurcation of the carotid artery are often referred to the maxillofacial surgeon. The differential diagnosis of these lesions includes

a variety of tumours among which carotid chemodectomas and vagus nerve schwannomas have a significant incidence. We describe a schwannoma originating from the vagus nerve mimicking a carotid body tumour, and compromising the function of the hypoglossal nerve. A diagnostic algorithm based on contemporary imaging modalities, which can be useful in order to distinguish between these two pathological entities is presented.

Conflict of interest The authors would like to declare that they have no conflict of interest.

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