A RARE CERVICAL MASS ASSOCIATED WITH PREOPERATIVE HORNER'S SYNDROME

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ABSTRACT

Introduction: *Schwannomas arising from the cervical sympathetic trunk are the most common benign nerve sheath tumors in the parapharyngeal space. They present as an asymptomatic slow-growing neck mass, without neurological symptoms. The purpose of this paper is to emphasize the rarity of the sympathetic chain cervical schwannoma with preoperative Horner's syndrome and aims to describe the presentation, imaging evaluations, surgical technique, and outcome.*

Case presentation*: A 51-year-old male was admitted in our department for a left neck mass associated with blepharoptosis, miosis and anhidrosis. The preoperative diagnosis of a sympathetic chain tumor relied on clinical and imaging studies. Horner's syndrome, which rarely appears preoperatively, was detected on preoperative clinical evaluation, contributing to a faster and more precise diagnosis. The internal carotid artery and internal jugular vein were altogether displaced antero-laterally on contrast-enhanced CT and MRI. The patient underwent surgical excision as the treatment of choice. Histopathological examination confirmed the diagnosis of sympathetic chain schwannoma. Postoperative the patient had first bite syndrome, pain in the left shoulder, voice hoarseness and coughing, foreign body sensation, tongue biting and deviation that gradually disappeared during the following 18 months.*

Conclusion*: In the case of a cervical mass, schwannoma must be considered as a diagnosis, particularly when there neurological signs are present. Preoperative diagnostic evaluation and rigorous planning of surgical intervention in cervical sympathetic chain schwannomas are essential. Our case supports the theory that a cervical schwannoma pushing forward both the internal jugular vein and the carotid artery suggests an origin from the sympathetic chain.*

Keywords: *cervical mass, cervical sympathetic chain, schwannoma, Horner's syndrome.*

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Introduction

Schwannomas are neuroectodermal benign nerve sheath tumors of peripheral nerves, originating in Schwann cells[1]. They are encapsulated benign tumors that rarely suffer malignant transformation into neurofibrosarcomas[2]. Surrounded by a distinct capsule, they are easily and successfully removed[3]. In the parapharyngeal space, schwannomas appear on the glossopharyngeal, vagus, accessory or hypoglossal nerve, with the second one being the most common. Schwannomas arising in the cervical sympathetic chain appear as elastic, well-defined neck masses, with a slow clinical course. While in the majority of cases the neural function is preserved, Horner's syndrome is not common on preop clinical examination[2]. Thus, the pre-operative diagnosis relies solely on imaging techniques. Multiparametric MRI is nowadays the standard investigation, with contrast-enhanced CT and ultrasound also appropriate in case MRI is not available. Magnetic resonance angiography or CT angiography of the major cervical vessels can easily differentiate schwannomas from carotid body tumors. Fine needle aspiration cy-

tology or biopsy is not recommended while imaging diagnosis can be accurate and sufficient[2,4].

In this paper we present a rare case of cervical sympathetic trunk schwannoma with preoperative Horner syndrome, making the diagnosis clear from clinical examination.

Case presentation

A 51 years old man presented with a tumor in the left cervical region with 2 years clinical course. Clinical examination revealed a 5 x 4 cm elastic, well defined, non-tender upper latero-cervical tumor and Horner's syndrome (blepharoptosis, miosis, anhidrosis) on the left side (Fig.1).

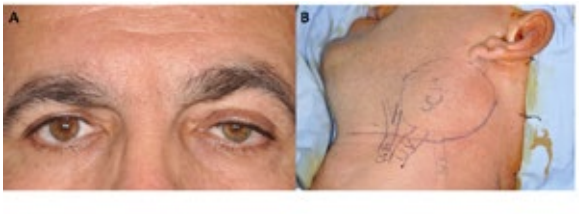

Fig. 1: Clinical examination and planning:
A. *Clinical image of pre-op Horner syndrome, presenting miosis and blepharoptosis*. ***B***. *Transcervical approach: SCH= schwannoma; CCA= common carotid artery; IJV= internal jugular vein; EJV= external jugular vein.*

An initial diagnosis of neural tumor was possible as there were signs for sympathetic chain involvement (Horner's syndrome).

MRI of the neck showed a heterogeneously contrast-enhanced mass, 5,5 cm in cranio-caudal direction, located in the vascular compartment of the left parapharyngeal space (retro-styloid), displacing both the internal jugular vein and the carotid artery anteriorly. As they were close together, the vagus nerve schwannoma was excluded from diagnosis (Fig. 2A).

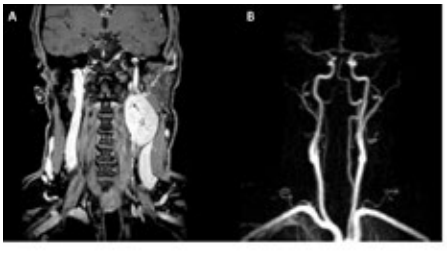

Fig. 2: Preop-imaging:
A. *Coronal MRI section defining the cranio-caudal extent of the lesion*. ***B***. *Magnetic resonance angiography of the major vessels revealing no hypervascularization or splaying of the internal and external carotid arteries, excluding a carotid body tumor.*

Magnetic resonance angiography of the major vessels revealed no hypervascularization or splaying of the internal and external carotid arteries, excluding a carotid body tumor. (Fig. 2B). Thus, the imaging diagnosis confirmed the clinical one, as cervical sympathetic trunk schwannoma.

Tumor was excised through a transcervical approach (Fig. 1B). The common carotid artery and the internal jugular vein were found to be displaced anteriorly, tumor originating in the sympathetic chain, leaving vagus, spinal, hypoglossal, glossopharyngeal or lingual nerves intact (Fig. 3A). The mass presented a well-defined capsule. Complete surgical excision was possible (Fig. 3B).

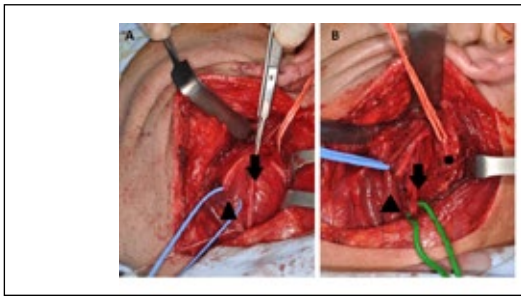

Fig. 3: Intra-operative aspects:
A. *Before tumor removal: arrow= vagus nerve; arrowhead= internal jugular vein*. ***B***. *After tumor removal: arrow= vagus nerve; arrowhead= internal jugular vein; circle= spinal nerve*

Macroscopically, the tumor presented as a whitish encapsulated nodule with focal areas of hemorrhage and frostbite appearance. A monomorphic cell proliferation - with elongated, fusiform cells presenting eosinophilic, reduced cytoplasm and hyperchromic, oval nuclei - was revealed. The cells were arranged in bundles that intersect and form hypercellular areas (Antoni A) alternating with hypocellular areas (Antoni B). At the level of the hypercellular areas, the nuclei palisaded and formed Verocay bodies.

At 1-month follow-up, beside the Horner syndrome, patient presented pain in the parotid area, initiated with each meal first bite. Shoulder syndrome on the left side was also present, along with voice hoarseness, coughing and foreign body sensation during deglutition. The patient reported tongue biting and deviation of the tongue to the left side.

At 6 months follow-up there was neither shoulder pain, nor tongue biting. Coughing was reduced. There was still first bite pain, voice hoarseness, foreign body sensation and Horner syndrome.

At 18 months follow-up the patient presented with Horner syndrome, but all the other symptoms disappeared.

Discussion

Cervical sympathetic chain schwannomas don't usually present with specific symptoms. MRI, CT or other paraclinical examinations cannot expose the nerve from which the tumor originated[5]. Radiological investigations can only limit the diagnosis to tumors originating in the retro-styloid compartment of the parapharyngeal space[2], ruling out salivary gland tumors or lymphadenopathies that originate in the pre-styloid compartment[6].

A clinical diagnosis is quite difficult to make and imaging studies are compulsory. In our case, MRI revealed an encapsulated tumor with low signal intensity on T1 and high signal intensity on T2-weighted images. The mass was located in the vascular compartment of the parapharyngeal space, excluding other tumors with origin in the pre-styloid compartment. Carotid artery was displaced anteriorly, suggesting a schwannoma originating either from the sympathetic chain or vagus nerve. While the internal jugular vein was not separated from the carotid artery the origin of the tumor was considered to be in the cervical sympathetic trunk. The presence of Horner's syndrome at clinical examination confirmed the diagnosis of sympathetic chain schwannoma.

In most cases, clinical and imaging studies confirm the diagnosis. Incisional biopsy is not needed and presents some risk. Ultrasound guided fine needle aspiration cytology, which would be less risky, provides minor diagnostic value in compact neural tumors[4]. Surgical excision and histological examination of the specimen provides the final diagnosis[9].

Complete excision is the treatment of choice. It is superior to enucleation as a curative procedure. Enucleation exposes to a low but not neglectable risk of local recurrence[11]. The main argument favoring enucleation is the possibility of sparing the nerve's fibers, but this advantage does not stand in this case where the Horner's syndrome was present on admission. Besides Horner syndrome, others postoperative complications were linked to the dissection of the parapharyngeal space and the nerves located in the area (glossopharyngeal, hypoglossal, vagus and spinal).

The patient was diagnosed with first bite syndrome. This was due to the denervation of sympathetic receptors that provoked a hypersensitivity of the parasympathetic receptors resulting in a supramaximal contractile response at the first bite of each meal.

The patient also presented pain in the left shoulder due to the dissection of the XI nerve. Because of the dissection of the vagus nerve the patient experienced voice hoarseness, coughing and foreign body sensation during deglutition. The patient reported tongue biting and deviation of the tongue to the left side (XII nerve dissection). In the case of a cervical mass, schwannoma must be considered as a diagnosis, particularly when there are neurological signs present. Preoperative diagnostic evaluation and rigorous planning of surgical intervention in cervical sympathetic chain schwannomas are essential. Our case supports the theory that a cervical schwannoma pushing forward both the internal jugular vein and the carotid artery suggests an origin from the sympathetic chain.

The rarity of the sympathetic chain cervical schwannoma with preoperative Horner's syndrome makes our case interesting to report.

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