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Caso Clínico

Complications of sympathetic chain schwannomatosis: a case report

Complicaciones de la schwannomatosis de la cadena simpática cervical: caso clínico

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Abstract

Objective: to report a case of cervical sympathetic chain schwannomatosis.

Study design: case report.

Introduction: Schwannomas are benign tumors arising from the Schwann cells of the peripheral nerves. Its occurrence within the cervical sympathetic chain schwannomas is rare, and it merges within the parapharyngeal space usually as an asymptomatic and solitary mass.

Case: a 42-years old female with a right parapharyngeal mass. An angio-magnetic resonance image revealed a tumor that slightly displaced of the internal carotid artery. Tumor excision was performed through a cervical approach. After surgery the patient developed a Horner' syndrome and a First-bite syndrome, the second successfully treated with a botulinum toxin injection.

Conclusion: sequelae derived from the excision of cervical sympathetic chain schwannomas include potential post-operative neural deficits as the Horner´s syndrome, and neuropathic pain as the first bite syndrome. We recommend the patient counseling about the potentials complications and sequelae of the intervention before the surgery is performed.

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Keywords: schwannoma, sympathetic denervation, Horner syndrome, postoperative pain.

Resumen

Objetivo: Presentar un caso de schwannomatosis de la cadena simpática cervical.

Diseño del estudio: caso clínico.

Introducción: los schwannomas son tumores benignos derivados de las células de Schwann de los nervios periféricos, su aparición en la cadena simpática cervical es rara, surgiendo dentro del espacio parafaríngeo usualmente como una masa asintomática.

Caso clínico: mujer de 42 años con una masa en espacio parafaríngeo derecho. Una angio-resonancia magnética un tumor que discretamente desplaza la arteria carótida interna. Se realiza la exéresis del tumor mediante un abordaje transcervical y tras la cirugía la paciente desarrolla un síndrome de Horner y un síndrome del primer mordisco tras la exéresis del tumor, éste último tratado eficazmente con la inyección de toxina botulínica.

Conclusión: las secuelas derivadas de la exéresis de los schwannomas de la cadena simpática cervical incluyen el déficit neurológico iatrogénico, como el síndrome de Horner, además de dolor post-quirúrgico neuropático, como el síndrome del primer mordisco. Nosotros recomendamos orientar e informar a los paciente de las posibles complicaciones y secuelas de la exéresis de estos tumores previo a la realización de la cirugía.

Palabras clave: schwannoma, denervación simpática, síndrome de Horner, dolor postoperatorio.

Introduction

Schwannomas are benign nerve sheath tumors derived from the Schwann cells of the peripheral nerves[1]. Approximately 20-45% of these tumors are situated within the head and neck region. At the parapharyngeal space, schwannomas can arise from the last four cranial nerves or the autonomic nerves[2]. Cervical sympathetic chain schwannomas are very rare and usually present as an asymptomatic and solitary cervical mass. Symptoms develop due to tumor growth and expansion[3]. Surgical resection is the treatment of choice but is not always recommended due to the potential post-operative neural deficits[2–4], thus, less invasive options should be considered. We recommend the patient counseling before surgery.

Case report

A 42 years-old female referred to our Department because of a right-side parapharyngeal mass. She had no history of chronic diseases nor surgeries, but she was a smoker of 7 cigarettes per day for the last 10 years.

On the physical examination, we observed a parapharyngeal mass that displaced the right tonsil to the

middle line, but no mass could be felt with the cervical palpation. A magnetic resonance image (MRI) and an angio-MRI revealed a well-delimited heterogeneous mass of 4.8 x 2.5 x 4 cm, with a heterogeneous contrast-enhancement and no association of cervical ganglia. The tumor did not affect the carotid bifurcation, but there was a slight displacement of both internal and external carotid arteries (Figure 1). The fine-needle aspiration cytology FNAC revealed a schwannoma (Figure 2).

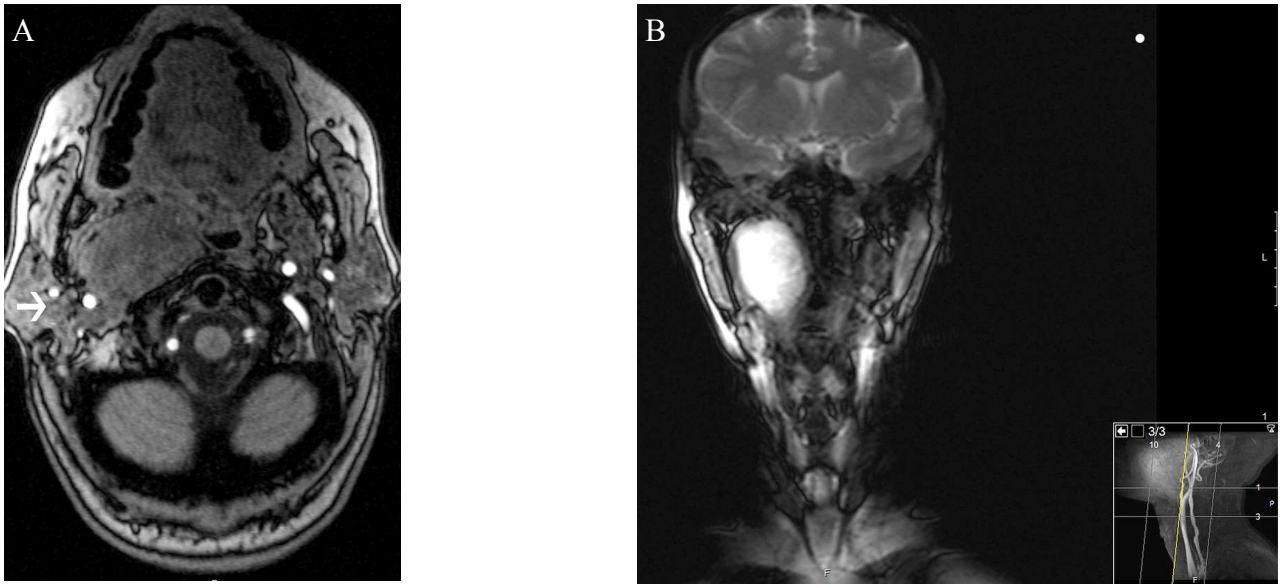

Figure 1a: MRI showing lateral and posterior displacement of both the internal and external carotid arteries (white arrow). Figure 1b: Angio-MRI showing no vascular involvement of the tumor.

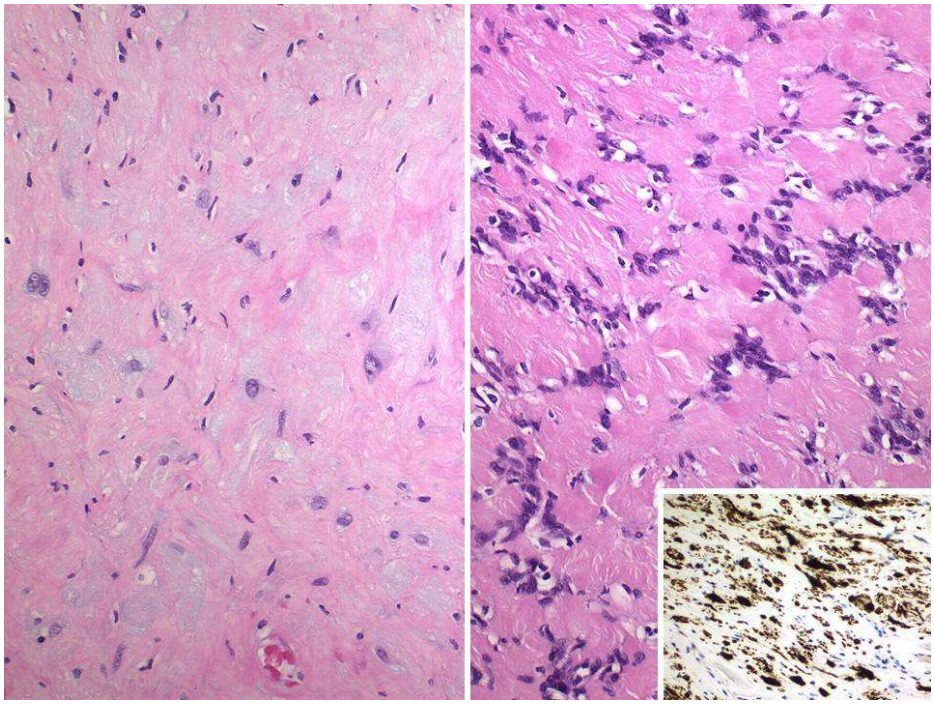

Figure 2: Fine-needle aspiration cytology: Schwannoma: the microscopic image on the left (hematoxylin -eosin) shows discrete nuclear atypia and mitosis figures. In the photograph on the right (hematoxylin-eosin) there are numerous palisaded nuclei (Verocay bodies). The immunohistochemical study was positive for the S100 protein (inset).

Surgical resection was performed through a transcervical approach. A thorough intervention was executed, identifying meticulously the cranial nerves ten and eleven, as well as the internal jugular vein, the main carroty artery and its bifurcation. We observed a well-defined tumor emerging posteriorly to the carotid sheath, at the bifurcation level (Figure 3), but the nerve of origin could not be identified. Immediately after surgery, the patient developed a right Horner´s syndrome and an ipsilateral vocal fold paresis. The vocal fold movement was recovered after 3 weeks, but the Horner´s syndrome remained, therefore the cervical sympathetic chain was determined as the origin. Four weeks after surgery the patient complained of an excruciating pain on the right preauricular area and ipsilateral mandible, triggered by eating. The diagnosis of a first-bite syndrome was made. The patient was given a first-line treatment with non-steroids anti-inflammatories (NSAI) and Gabapentin with no improvement; therefore, we applied an intraparotid injection of 75 units of type A botulinum toxin. The patient reported improvement after 24 hours of the injection.

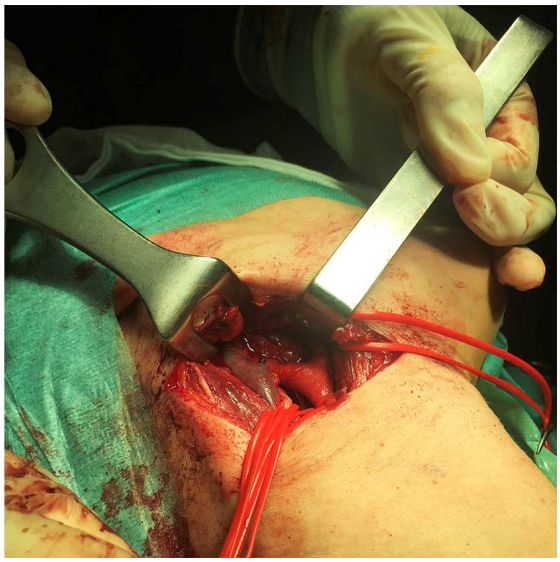

Figure 3: Tumor emerging posteriorly to the carotid sheath, at the carotid bifurcation level. During the surgery the main carotid artery, the internal jugular vain, the hypoglossal nerve and the vagus nerve were protected with vessel-loops.

Discussion

Although schwannomas constituted the most common nerve sheath tumors, they are, nevertheless, very rare. At the parapharyngeal space, the vagus nerve is the most common nerve of origin[4]; followed by the cervical sympathetic chain, but, within this compartment schwannomas can also arise from cranial nerve IX, XI or XII . The time of presentation is usually between the 2nd and 5th decade of live, and males and females are equally affected.

Cervical sympathetic chain schwannomas (CSCS) usually present as an asymptomatic and solitary cervical mass. Symptoms may develop due to tumor growth and expansion, so patients can indicate dysphagia

or odynophagia as associated symptoms[3]. Expansion of the tumor usually occurs medially from the lateral pharyngeal wall, which displace the tonsil and the soft palate to the middle line, as in our patient. This vague presentation makes it difficult to determine the diagnosis. Differential diagnosis should be made with other parapharyngeal space tumors, specially paragangliomas of the carotid body or glomus jugulars tumors; as well as pleomorphic adenoma, which is the most common tumor arising from this compartment.
Radiographic imaging is a key step in the diagnosis of the CSCS. Findings on the computer tomography (CT) can be helpful evaluating the tumor size and location, nevertheless, MRI is essential, as it can help indicate the nerve of origin, based in the tumor relationship with the great vessels[4,5] . The CSCS can cause the anterior displacement of main the carotid artery and jugular vein together or cause the separation between the internal and externa carotid arteries, as the CSC arises behind the carotid sheath. Vagus nerve schwannomas, on the other hand, cause separation between the carotid artery and the internal jugular vein. Another crucial differential diagnosis should be made with paragangliomas, which exhibit an extreme bright contrast enhancement with a typical ´´salt and pepper pattern´´ in MRI[6].
Fine-needle aspiration biopsy is the preferred method for diagnosis of a neck mass[7], but it offers little information in the diagnosis of CSCS[8] due to the dense stromal components of these tumors or cystic degeneration within them[8]. When the aspirate provides enough cellular material the histology shows fusiform cells with elongated nuclei that are grouped in areas of high cellularity, called tissue type A of Antoni and in other laxer areas, of lower cellularity (Antoni type B). In the Antoni A type areas, the cells can be organized in the form of a palisade forming the so-called Verocay bodies, characteristic of this type of tumors but not pathognomonic. Immunohistochemically these tumors are positive for protein S-100[9].
While schwannomas have a slow growing rate, low recurrence rate and a malignant transformation is very rare, when arising from the parapharyngeal space treatment is mandatory because of the mass effect.
The surgical intervention can be challenging because it´s location and relationship with the great vessels and lower cranial nerves within the parapharyngeal space[10].
The management of these tumors is not clearly standardized[3], as surgical resection may be the treatment of choice, but is not always recommended due to the potential post-operative neural deficits associated with it[2], thus, less invasive options, like volumetric growth observation by radiological images[3], should be considered. Therefore, we recommend the patient counseling before surgery. Horner´s syndrome is to be expected after surgery, which appears inevitability, but it can appear pre-operative due to nerve damage caused by the tumor. Usually patients remain asymptomatic, except for visual accommodation issues.
On the other hand, the first bite syndrome (FBS) is a well described but rare complication of the parapharyngeal space surgery, characterized by presence of severe pain in the parotid region initiated with the first bite of each meal[11]. It can also appear after surgery of the infratemporal fossa and the deep parotid space due to sympathetic trunk lesion which leads to autonomic imbalance. The parotid myoepithelial cells receive innervation from both sympathetic and parasympathetic systems, so the denervation of sympathetic receptors located on these cells provoke a hypersensitivity of the parasympathetic receptors resulting in a supramaximal contractile response at the first bite of each meal[12,13]. The pain usually starts to appear several days after surgery and it tends to disappear within the first year. Pain can be so exquisite that patients

avoid eating in order to prevent pain. The injection of botulinum toxin into the parotid gland has shown to diminish the intensity of pain.

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