



PARAPHARYNGEAL SCHAWANNOMA. CASE REPORT

SCHWANOMA PARAFARINGEO

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ABSTRACT

Parapharyngeal tumor lesions present a low incidence, representing between 0,5 and 0,8% of all head and neck tumors. Approximately 80% show benign behavior. The uniqueness of these lesions derives from their complex anatomical situation and the symptoms with which they usually appear, being in most cases nonspecific and almost always derived from the compressive effect produced by the lesion on the oropharynx and the oropharynx. Schwannomas of the parapharyngeal space are very rare tumors that originate from the sheath of schwann, generally slow growing, and are usually asymptomatic. Treatment is surgical, often complex due to the anatomical location. The case of a 42-year-old female patient with a diagnosis of Schwannoma in the left parapharyngeal region, demonstrated by incisional biopsy, is presented; undergoing surgical treatment, which was carried out without complications.

Key words: Parapharyngeal space; Schwannoma (source: MeSH NLM).

RESUMEN

Las lesiones tumorales de localización parafaríngea presentan una escasa incidencia, representando entre el 0,5 y el 0,8% de todas las tumoraciones de cabeza y cuello. Aproximadamente el 80% presentan un comportamiento benigno. La singularidad de dichas lesiones deriva de su compleja situación anatómica y de la clínica con la que suelen debutar, siendo en la mayoría de los casos inespecífica y casi siempre derivada del efecto compresivo producido por la lesión sobre la oro y rinofaringe. Los Schwannoma del espacio parafaríngeo son tumores muy infrecuentes, que se originan de la vaina de schwann, generalmente de crecimiento lento, suelen ser asintomáticos. El tratamiento es quirúrgico, muchas veces complejo por la localización anatómica. Se presenta el caso de una paciente femenina de 42 años de edad con diagnóstico de Schwannoma en la región parafaríngea izquierda demostrada mediante biopsia incisional; realizándosele tratamiento quirúrgico, el cual se llevó a cabo sin complicaciones.

Palabras clave: Espacio parafaríngeo; Schwannoma (fuente: DeCS BIREME).

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INTRODUCTION

Schwannomas are the second most frequent tumors located in the parapharyngeal space. Although most of these tumors are found in the head and neck, those with an extracranial location only constitute 10% of the total⁽¹⁾. Parapharyngeal tumor lesions present a low incidence, representing between 0.5 and 0.8% of all head and neck tumors^(1,2). Approximately 80% show benign behavior⁽²⁻⁴⁾. The uniqueness of these lesions derives from their complex anatomical situation and the symptoms with which they usually appear, being in most cases nonspecific and almost always derived from the compressive effect produced by the lesion on the oropharynx and rhinopharynx, presenting symptoms such as dysphonia, compressive pain or pharyngeal mass. Secretory paraganglioma, although uncommon, can cause hypertension, sweating, nausea, palpitations or headache⁽³⁾. These are usually tumors that affect adults, without differences between the sexes and are mostly benign.

The pathological study of the tumor reveals a well encapsulated mass made up of spindle cells type A of Anoni and type B of Antoni, the immunohistochemistry shows an intense expression of the S-100 protein⁽⁵⁾.

The prognosis for a person with schwannoma is excellent when the mass has been completely removed. Local recurrence is infrequent, being related to incomplete exegesis of the neoplasm^(6,7).

CLINICAL CASE

42-year-old female patient with a 20-year history of migraine. She presented to the Internal Medicine service of the Hospital Edgardo Rebagliati Martins in Lima, Peru, reporting a 2 months disease of progressive onset and characterized by hemicranial

and left mandibular headache, of moderate intensity, accompanied by increased volume in the cervical region on the same side; also, mild odynophagia and dysphonia. Physical examination revealed a mass of approximately 5 cm in diameter in the left maxillary region, not adhered to deep planes, with non-hyperthermic surrounding skin and no color changes, nor fixed to deep planes, on bimanual manipulation it was discreetly painful on palpation. shallow and deep. Given the suspicion of an expansive parapharyngeal process, an imaging study was carried out using a Cervical Tomography with contrast (Figura 1), where a large solid left parapharyngeal mass with heterogeneous contrast enhancement and areas of peripheral calcification measured TD 38mm DL 50mm DBH 40mm, and that extends cranially towards the jugular foramen and displaces the cervical vascular structures anteriorly, without infiltration; causes mass effect with airway compression. In addition, a nuclear magnetic resonance study was performed (Figura 2, Figura 3), where it shows a solid image with well-defined edges, an ovoid shape in the left parapharyngeal space, exerting an expansive effect and moving the midline from left to right. Intramedullary extension is not observed. Subsequently, a BAAF was performed with ultrasound guidance of the lesion, the result of which was not conclusive for the diagnosis, which is why the case was reassessed, deciding to take an incisional biopsy by the Head and Neck Surgery Service, which was sent to histopathological study; Schwannoma was reported, being transferred for surgical treatment

definitive; A cervical approach was used (Figura 4, Figura 5), and the surgical specimen was sent for histopathological study, the result of which confirmed our initial diagnosis: Parapharyngeal Schwannoma.

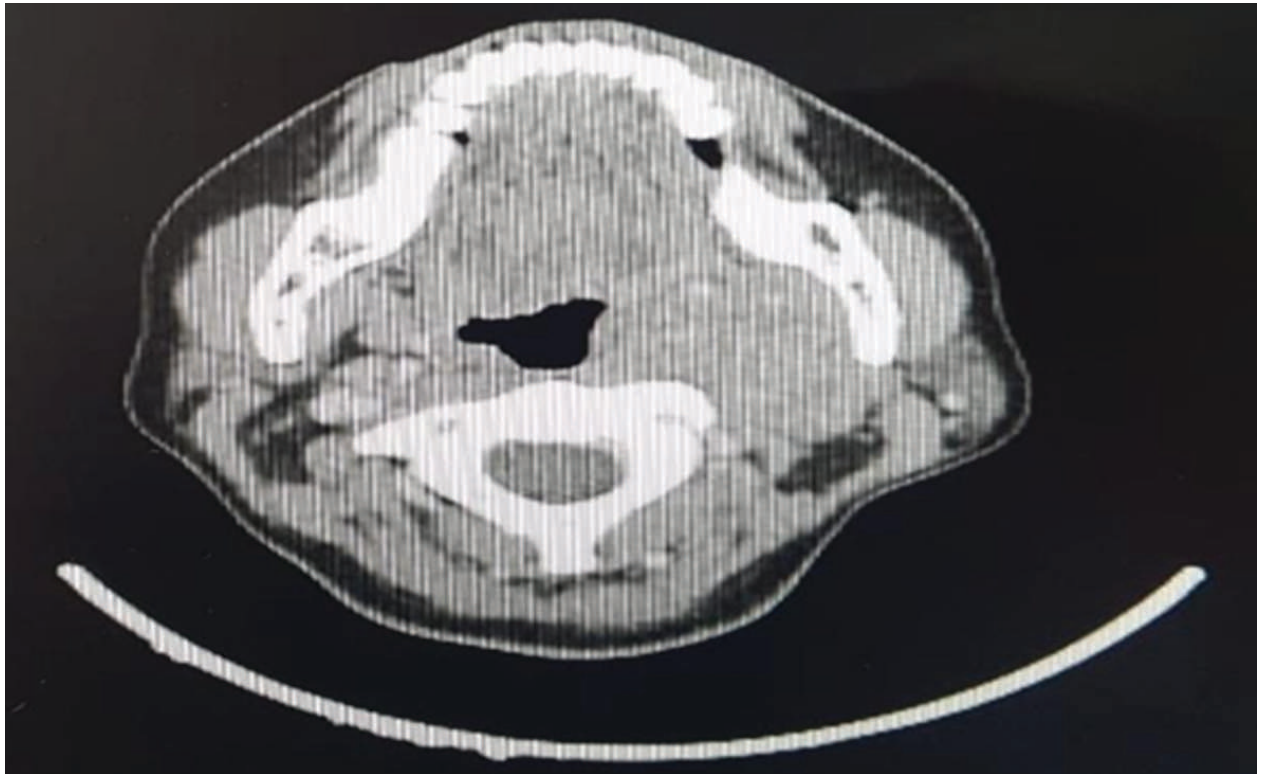


Figure 1. Contrast Tomography Scan of the neck, a large solid left parapharyngeal mass is evidenced with heterogeneous contrast enhancement and areas of peripheral calcification, measures DT 38mm DL 50mm DBH 40mm, which extends cranially towards the jugular foramen and displaces the cervical vascular structures anteriorly, without infiltration; causes mass effect with airway compression.

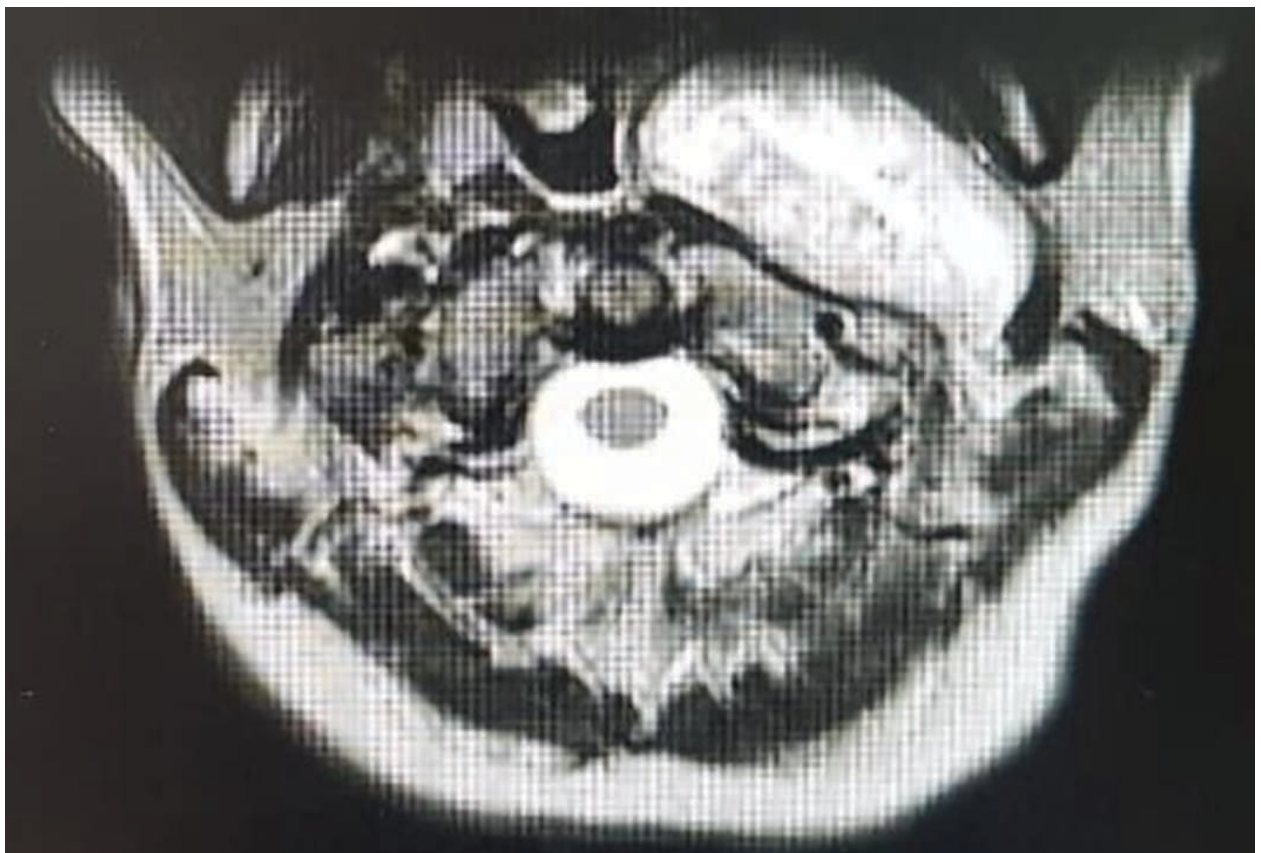


Figure 2. Magnetic Resonance Imaging of the neck with contrast, shows a solid image with well-defined edges, an ovoid shape in the left parapharyngeal space, exerting an occupational effect and shifting the midline from left to right. Intramedullary extension is not observed.

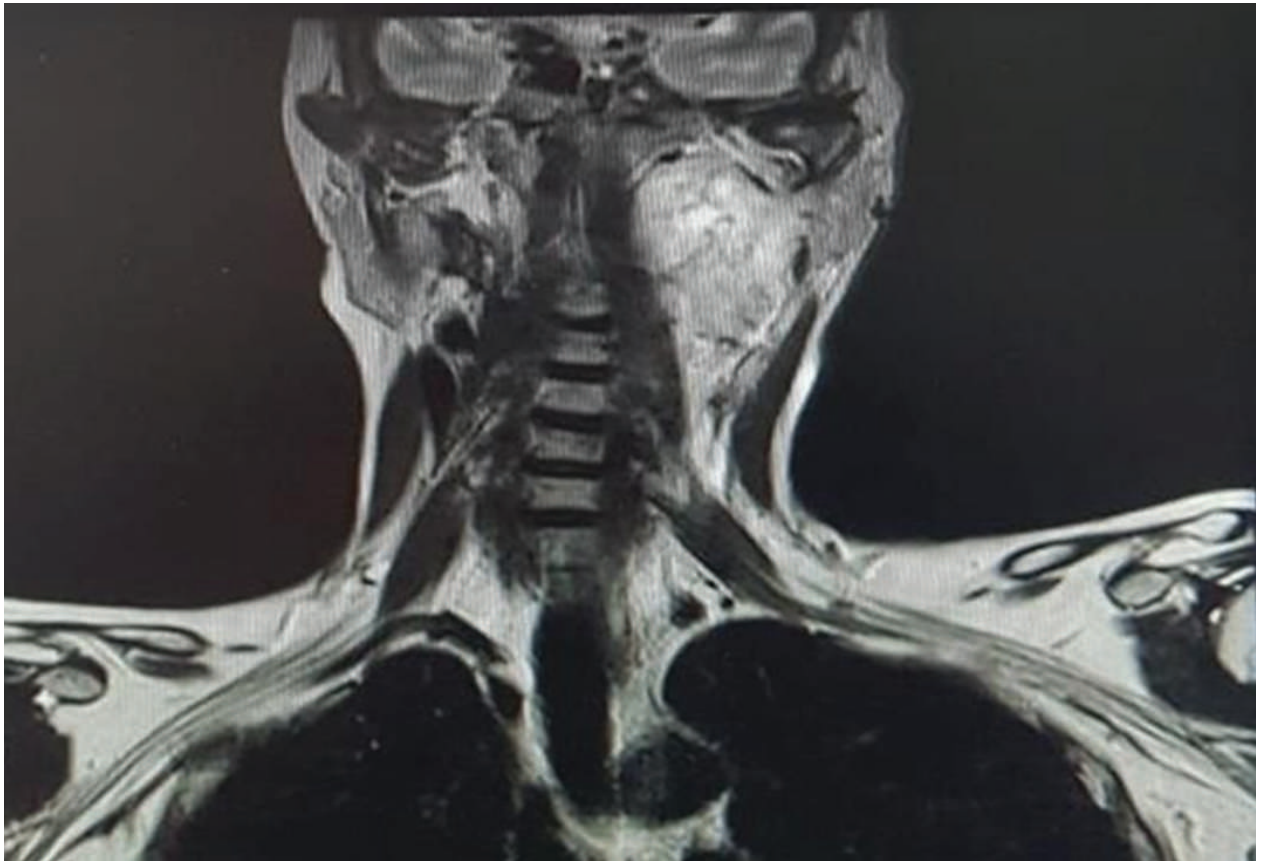


Figure 3. Taking an incisional biopsy, a cervical approach was used by the Head and Neck Surgery Service.

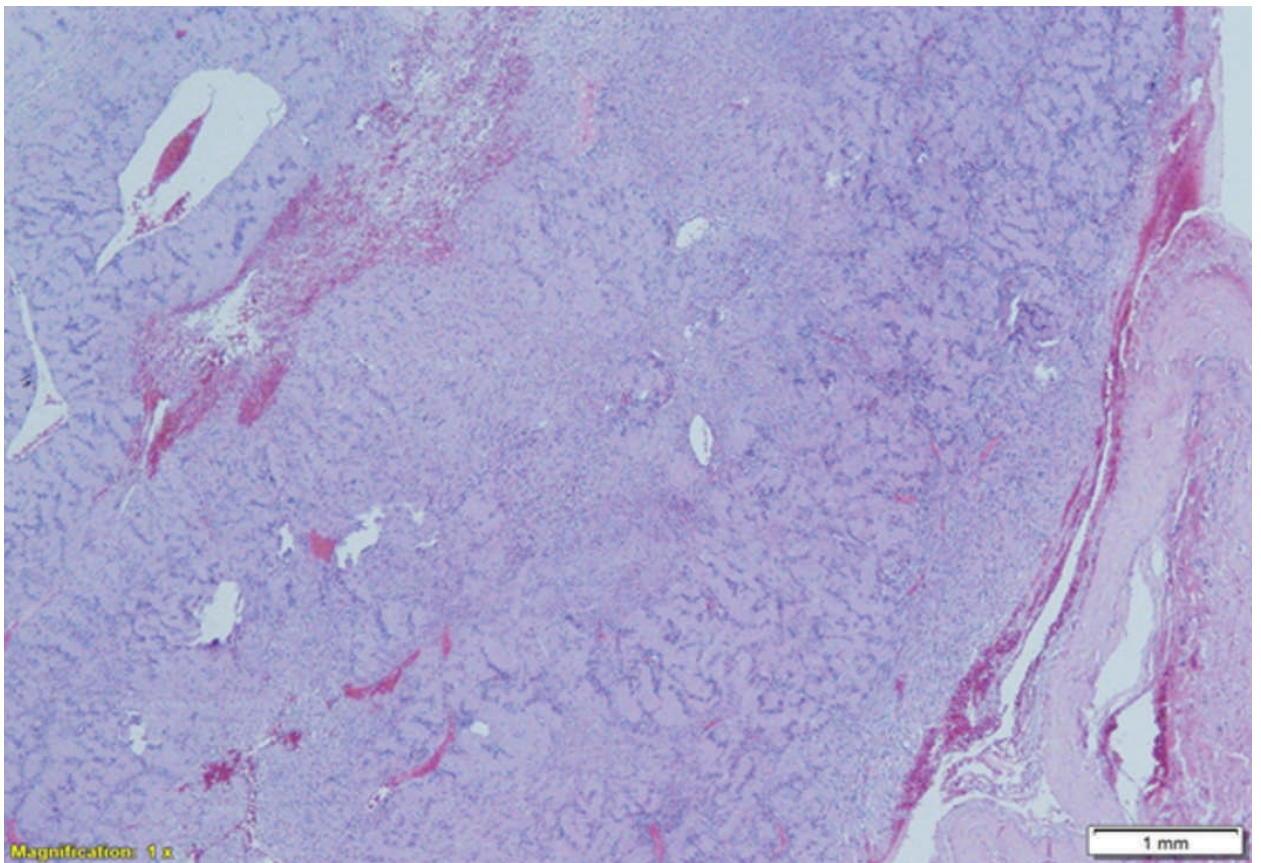


Figure 4. At lower magnification, a well-delimited and encapsulated tumor can be seen, and at this magnification the presence of palisade-shaped nuclei (Verocay bodies) can be seen.

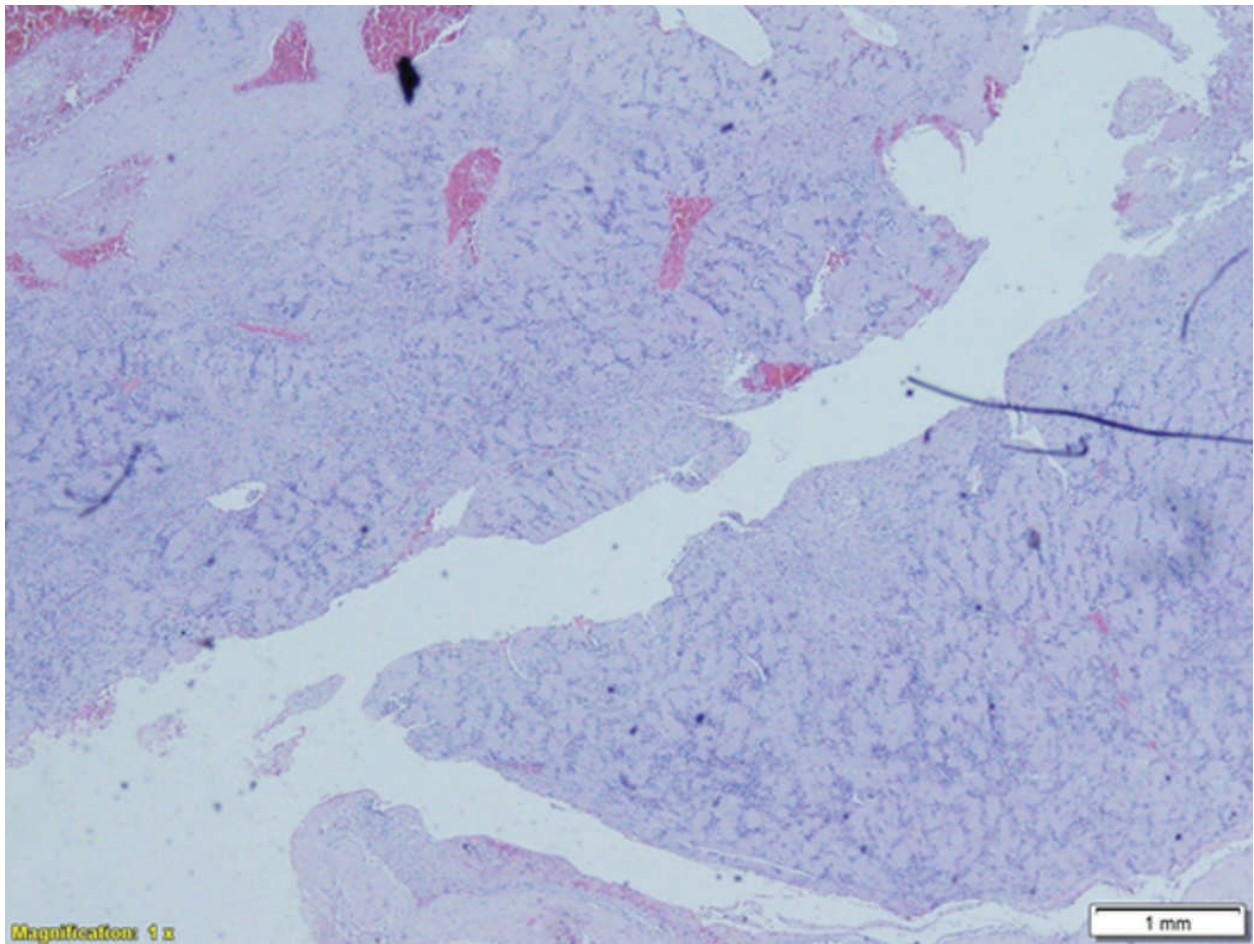


Figure 5. In addition to the Verocay bodies, vascular dilatation is seen, another frequent finding in Schwannomas.

DISCUSSION

Parapharyngeal space tumors account for 0.5% of all head and neck neoplasms⁽²⁾. Most of the lesions located in this space are benign primary tumors that originate from the vagus nerve, the most common being the schwannoma. Schwannoma is the most common histologic subtype of vagus nerve tumor after paraganglioma. It usually appears between the third and sixth decades of life, being more frequent in young adults⁽⁵⁾, presenting a similar frequency in both sexes⁽³⁾. Clinically without symptoms, it is diagnosed as a unilateral laterocervical mass mobile only in a solitary lateral direction, except in Von Recklinghausen's disease^(6,8) and in a rare syndrome known as schwannomatosis⁽²⁴⁾ where the presentation is multiple. They can be palpated as masses on the medial border of the sternocleidomastoid muscle. Its expansive growth through the parapharyngeal space can make it go unnoticed to a large size. If its size is considerable, it can cause very nonspecific signs such as pharyngeal paresthesia, cough, odynophagia due to pharyngeal

compression, dysphonia. A characteristic, although not always present, clinical sign of cervical vagal schwannoma is paroxysmal cough on palpation of the tumor and even serous otitis^(5,6,9). Symptoms derived from compression of nerve structures IX, X, XI, XII, of the sympathetic will be seen in cases of large neurinomas, or in malignant tumors and chemodectomas. Pain, trismus, and cranial nerve lesions suggest malignancy.

The diagnosis will be based on the clinic, the physical examination, being important the bimanual palpation, which allows to recognize the continuity of the tumor and determine its origin. Oropharyngeal and oral cavity visualization is important to rule out other primaries in these locations that could be producing secondary metastatic involvement in the parapharyngeal space. A pulsatile cervical mass may indicate the presence of a vascular tumor, such as a paraganglioma. The physical examination should include an examination of the cranial nerves and the performance of a flexible nasopharyngolaryngoscopy⁽²⁾.



The radiological study using Computed Tomography (CT) and Magnetic Resonance (MRI) will be of vital importance. On CT, they are tumors with a density similar to the muscle, more less rounded and well defined with a medial and lateral fatty plane, as their most frequent origin is the X pair or the sympathetic chain, they usually displace the internal carotid artery anteromedially. MRI provides excellent information in the study of soft tissues, being very useful in this type of tumors and that region, they are hypointense tumors on T1 and hyperintense on T2 and on T1 with gadolinium. Differential diagnosis should be made with carotid artery aneurysm, with neurogenic tumors of cranial nerves IX, X, XI and XII or of the cervical sympathetic plexus⁽¹⁴⁾, with paraganglioma, meningioma, lymphoma and teratoma⁽¹⁵⁾ However, the majority of lesions involving the retrostyloid parapharyngeal space are peripheral nerve sheath tumors, head and neck paragangliomas, or metastatic lymphadenopathy⁽²⁴⁾.

The performance of FNAB in tumors of the parapharynx can reach 86% - 99% especially in lymphoma, primary malignant tumors or metastases of malignant tumors^(22,24). However, FNAB would not be helpful in distinguishing benign from malignant paragangliomas or benign from malignant peripheral nerve tumors. As in our case, it was not a contribution for diagnosis.

The macroscopic examination usually corresponds to

a white, yellowish, well-defined mass with a firm and elastic consistency, sometimes with cystic areas. being typical the presence of necrosis, hemorrhage and intratumoral cystic degeneration⁽²³⁾. From the microscopic point of view there are two groups: Antoni A, a compact cellular composition in palisade and Antoni B, formed by a loose structure characterized by myxoid degeneration, although most of the time they are associated, while others defend that hemorrhage it is responsible for vacuolar degeneration due to hemosiderin deposition. Immunohistochemical techniques show an intense expression of the S-100 protein in these tumors, an antigen of the tissues derived from the neural crest^(20, 21).

The treatment is surgical, although due to the infrequent presentation of tumors in this location it is difficult to obtain surgical experience, there are different approaches, which will depend on the size and location of the tumor, the most used being the cervical route, suitable for most tumors, in larger cases or for tumors of pre-styloid location, the cervicoparotid approach can be useful. The most frequent complications in surgery are injury to the nerve itself and its resulting deficit. Recurrences are rare and are usually the result of incomplete excision, generally having a good postsurgical prognosis. Regarding the prognosis, in cases of benign tumors, when a complete resection is achieved, the risk of recurrence should be low to null.

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