CLINICAL CASE
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Palabras clave:
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ABSTRACT

Schwannoma or neurilemoma is known as a rare or frequent tumor arising from Schwann cells (glial cells found in the peripheral nervous system) and, in most situations, behaves benignly. Our case illustrates a rare pathology, a benign tumor that differentiates along Schwannian lines. Some of these tumors may have non-negligible mitotic activity, but their behavior is benign; however, these types usually occur in the deep soft tissues and rarely involve the subcutaneous tissue. During this report, we will deal with the case of a patient in her third decade of life who presents a cervical tumor with growth during three years until the medical-surgical approach, after the onset of symptoms, although without data of airway compromise, hemodynamic status or neurological alterations, but presenting significant pain and difficulty in cervical mobilization. It is complemented by imaging studies that suggest ruling out branchial cysts versus pleomorphic adenoma. The treatment of choice is excision since usually the schwannoma displaces and compresses the residual nerve in its periphery and can be removed without affecting the nerve, in this case, explicitly speaking of direct involvement of the left hemilateral vagus nerve (X pair), the tumor was dissected and enucleated seeking to preserve the functions of the nerve mentioned above. Subsequently, the patient presented data of Horner's syndrome during the immediate and immediate postoperative period with ptosis and enophthalmos, lasting for two months with alterations that remitted favorably, without data of paralysis, alterations in swallowing or dysphonia, only continued with pain in the temporal region when chewing.

RESUMEN

El Schwannoma o neurilemoma es conocido como una tumoración poco común o frecuente, proveniente de las células de Schwann (células gliales que se encuentran en el sistema nervioso periférico) y en la mayoría de las situaciones se comportan de manera benigna. El caso que presentamos es ilustrativo de una patología poco común, una tumoración benigna que se diferencia a lo largo de las líneas schwannianas, algunas de estas tumoraciones pueden tener actividad mitótica no insignificante, pero su comportamiento es benigno; sin embargo, este tipo de tumores suelen aparecer en los tejidos blandos profundos y sólo raras veces comprometen el tejido subcutáneo. Durante este reporte abordaremos el caso de una paciente de la tercera década de la vida, la cual presenta una tumoración cervical con crecimiento durante tres años hasta el abordaje médico-quirúrgico, posterior a inicio de sintomatología, aunque sin datos de compromiso de vía aérea, estado hemodinámico o alteraciones neurológicas, pero sí presentando importante dolor y dificultad a la movilización cervical. Se complementa con estudios de imagen que sugieren descartar quiste branquial contra adenoma pleomorfo. El tratamiento de elección es la escisión, ya que habitualmente el schwannoma desplaza y comprime el nervio residual en su periferia, pudiendo ser retirado sin afectar al nervio, en este caso específicamente hablando de afección directa al nervio vago (X par) hemilateral izquierdo, se diseca y enuclea la tumoración con lo que se busca preservar las funciones del nervio ya mencionado. Posteriormente la paciente presenta datos de síndrome de Horner durante el postquirúrgico inmediato y mediato con ptosis y enoftalmos, llegando a durar con alteraciones que luego de dos meses remiten favorablemente, sin datos de parálisis, alteraciones en la deglución o disfonía, sólo continua con dolor en región temporal a la masticación.

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INTRODUCTION

Schwannoma, neurinoma, neurolemoma, or neurilemoma is the formation of a benign neoplasm frequently seen in association with cranial nerve VIII (vestibulocochlear) and neurofibromatosis II.

These tumors were first described by Virchow and Verocay in 1910, but it was not until 1932 that Masson coined the term "schwannoma".

It is a rare, asymptomatic tumor originating from Schwann cells near a peripheral nerve. It usually appears in the fourth and fifth decades of life, with no apparent distinction between sexes, being its location predominantly in the head, neck, and extremities, and of single appearance; multiple forms are usually associated with Von Recklinghausen's neurofibromatosis. According to Daly and Roesler, lateral tumors evolve from cutaneous or muscular branches of the cervical plexus or the brachial plexus. In contrast, medial tumors arise from the last four cranial nerves and the cervical sympathetic chain.[1]

It is a pathology with generally slow clinical evolution, thus causing a late diagnosis in most patients suffering from this pathology.

At the time of diagnosis, it is essential to perform a correct and complete anamnesis and, above all, to have high-resolution imaging studies as diagnostic support (computed tomography scan, magnetic resonance imaging, and carotid arteriography).

According to pathological anatomy, they are subcutaneous, encapsulated lesions characterized by a double histological pattern, the Antoni A and B areas.

The areas of Antoni A are made up of the cellular component of the lesion, characterized mainly by dense clusters of Schwann cells, whose nuclei are arranged in some areas in a palisade, giving rise to two parallel rows separated by the prolongations of the Schwann cells, characteristics known as Verocay bodies.

Areas of Antoni B tend to present hypocellularity with irregular formation and predominate in a loose myxoid stroma with blood vessels and chronic inflammatory cells.

Most are sporadic and solitary but may be associated with neurofibromatosis, especially in neurofibromatosis type 2 NF2 and schwannomatosis (a distinct entity).[2]

Of the schwannomas, 10% are extracranial, and of this, 25-45% occur in the head and neck; 50% of the parapharyngeal schwannomas have origin in the vagus nerve, with the cervical sympathetic chain being the second most frequent site of schwannoma settlement. According to Laconi and Faggioni, those of the cervical sympathetic chain are extremely rare, with less than 60 cases reported in the English literature.[2]

Surgical excision continues to be the therapeutic management for schwannomas; with a low recurrence rate; complementary treatment is not recommended. Sending the anatomopathological specimen for study is necessary to confirm the diagnosis of schwannoma.

PRESENTATION OF THE CASE

The case is a 21 years-old female patient from Ciudad Juarez, Chihuahua, Mexico; she is a catholic housewife with a high-school education and a history of a normal pregnancy, with eutocic delivery at term and binomial discharge. She has no other relevant history, no chronic degenerative diseases, or previous surgeries. She denies smoking, alcoholism, and drug addiction.

Her current condition started four years ago (2018-2021), with localized pain in left hemi collar and a progressive increase in volume, without data of dysphagia or dyspnea, which begins with gradual growth until causing pain, 3/10 in intensity according to visual analog pain scale (VAS) and difficult cervical mobilization. She had no infectious process, fever, or constitutional symptoms history.

Physical examination revealed the presence of a cervical tumor measuring approximately 12 × 8 × 10 cm, indurated, and fixed to deep planes, covering regions II, III, and Va of the left hemi collar *(Figure 1)*; it was slightly painful on palpation with a 3/10 intensity on VAS, without difficulty in opening the mouth, no facial asymmetry nor airway involvement.

There was no systemic inflammatory response syndrome, hemodynamic instability, or cardiopulmonary compromise. As a

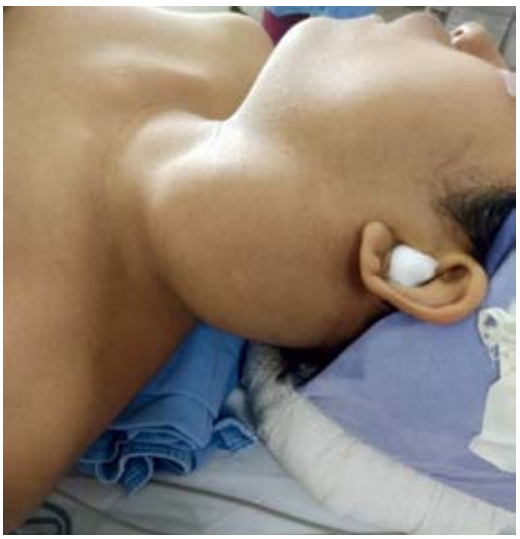

Figure 1: *Ovoid tumor in regions II, III, and Va of the left hemi collar.*

diagnostic complement, a CT scan is requested, which reports a large, rounded mass of solid appearance with regular edges, well delimited, heterogeneous, predominantly hypodense, with small areas of lower density inside, without identifying calcifications, with a density between 24 and 37 HU and that after the administration of intravenous contrast presents a density between 33 and 87 HU. It was in the left parapharyngeal level of 6.6 × 7.5 × 9.2 cm, as a left parapharyngeal space lesion with a suspected branchial cyst versus pleomorphic adenoma versus hemorrhagic branchial cyst *(Figures 2 to 4)*.

The study protocol was followed, and the surgical procedure was programmed. On the operating table with the patient in supine decubitus with Rossier position, a cervicotomy was performed through a Paul André cervical incision. After dissecting by planes (skin, subcutaneous cellular tissue, platysma), the sternocleidomastoid muscle was located, and a tumor located in the parapharyngeal space between the internal jugular vein and carotid artery *(Figure 5)* was found; it was dissected and enucleated, and a meticulous subcapsular dissection was performed trying to preserve the functions of the nerve of apparent origin, in this case, the X cranial nerve, to avoid its resection. The piece was sent to pathology; hemostasis was performed, and a negative pressure closed drainage of type Drenovac of 1/8 was placed. The surgical incision was closed by planes; in the fascia, Vicryl 2-0 was used, and the skin was approached with a nylon 3-0 intradermal stitch, and the surgical event ended.

During a four-day hospital stay, analgesic management and prophylactic antibiotic therapy were administered, and serous drainage decreased to report a minimum of 20 cm^3 in 24 hours; by then, it was removed. In the immediate postoperative course, ptosis and enophthalmos were seen, in addition to pain at the surgical site. They all improved and showed a clear progression. Therefore, the discharge was decided with outpatient follow-up at two weeks, one month, two months, and four months later, with remission of postoperative sequelae almost entirely, with no data of paralysis, swallowing disorders, or dysphonia. Currently, the patient only presents pain in the left lower jaw when chewing and pain in the ipsilateral temporal region.

Subsequently, a histopathological report was obtained with folio Q215-2022, where it was mentioned a neck tumor compatible with schwannoma, with an immunohistochemistry

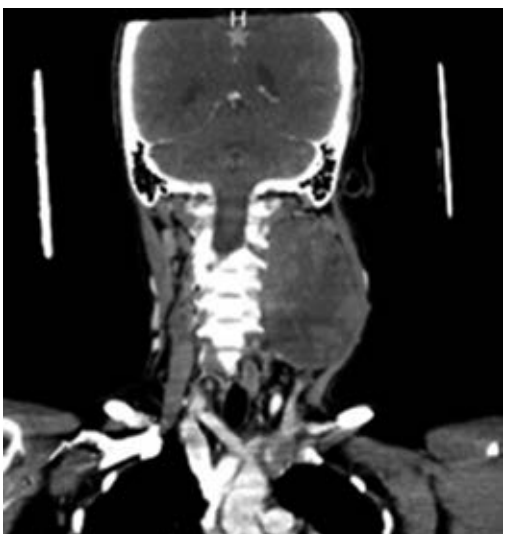

Figure 2: *Coronal section computed tomography scan showing a large mass with a solid appearance at the left parapharyngeal level.*

report with a result describing diffuse S100 (+++/+++) vimentin positive (+++/+++) *(Figures 6 and 7)*.

DISCUSSION

According to Araujo CE and Zhang H, this type of cervical tumor usually has a slow growth of approximately 2 to 3 mm per year;[3,4] according to the time of evolution of the reported case, if it complied with the above mentioned, it would measure no more than 1 cm; however, it measured ten times more. A factor is added because lesions of this type could have variable growth.

Luisa Gil and Marta Ortega Millán mention that one of the critical characteristics when interpreting imaging studies, given the fusiform morphology of the lesion with the major longitudinal axis, is that it follows the path of the nerve, which is characteristic in this type of lesion,[5] and shares the same characteristics in the report addressed.

It is of vital importance to know this pathology in the first instance to suspect it and to be able to make a diagnosis employing imaging studies. According to G. Cavallaro, fine needle puncture (FNA) has a low yield.[6]

The literature also mentions the high resistance of schwannomas to radiotherapy, thus leaving surgery as the treatment of choice. Precisely, the technique that consists of enucleation of these tumors, with preservation and protection of the nerve, total excision of the lesion of the neurovascular bundle is performed through a transcervical approach.[7]

According to Gibber, surgical resection can be achieved by enucleating the schwannoma of the vagus nerve and preserving it by microsurgical dissection and neuromonitoring,

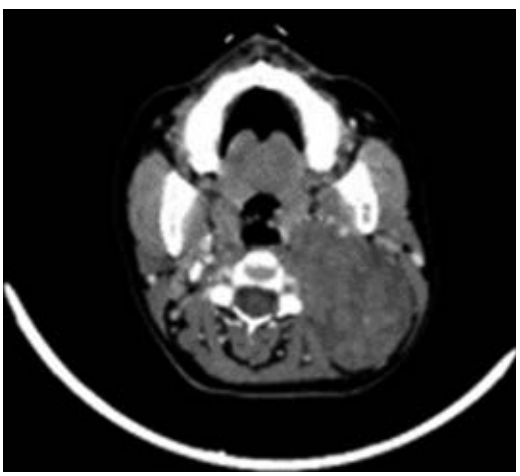

Figure 3: *Axial computed tomography scan with a solid appearing mass in left parapharyngeal space.*

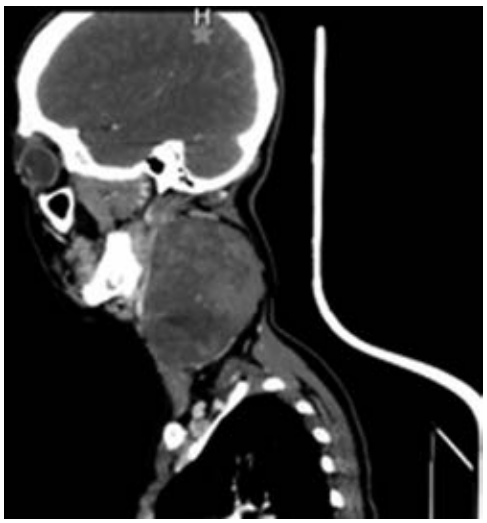

Figure 4: *Sagittal section of a computed tomography scan showing a solid lesion in the cervical region of approximately 10 × 9 cm.*

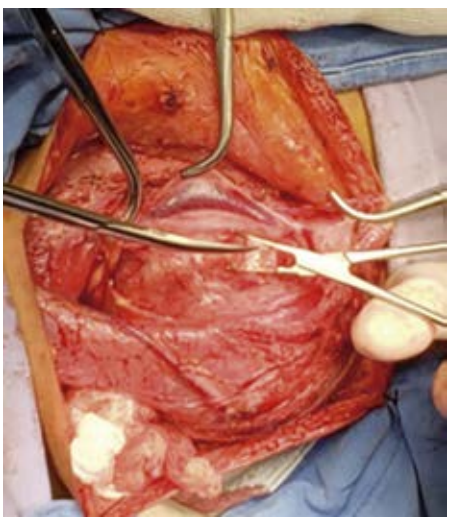

Figure 5: *Retraction of the internal jugular vein evidencing the origin of the schwannoma from the vagus nerve.*

thus reducing postoperative morbidity.[8] This is also mentioned by Kwok and Davis when referring to the use of intraoperative electrophysiological monitoring as a helpful tool during schwannoma resection.[9] In our particular case, the pathology approach was performed with high suspicion of a branchial cyst, so neuromonitoring was never considered as such, due to enucleation of the nerve from the schwannoma, preserving the nerve without monitoring.

As one of the complications during the follow-up at two and four months, the patient presented data of ptosis and enophthalmos, characteristic of Horner's syndrome, which, according to Massimo Politi and Faith Bingol, is one of the rare post-surgical manifestations with a duration of up to four months after the surgical event,[10,11] during which our patient had a good evolution and progression, reducing the clinical picture almost entirely. The patient presented with pain in the wound area and swallowing disorder with functional limits, with no data of paralysis or dysphonia.

CONCLUSIONS

Schwannoma is an infrequent pathology within neck tumors, but it is essential to consider it in soft tissue tumor pathology. Its approach is clinical, and with the support of imaging studies to reach the suspicion, better options for a trans-surgical approach, such as neuromonitoring, are considered, which offers a wide area of opportunity where professionals can obtain better results.

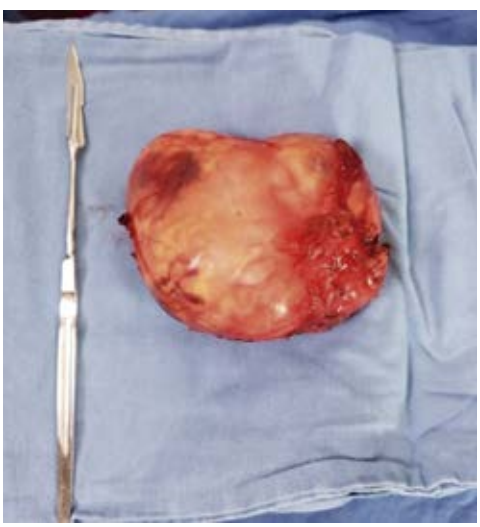

Figure 6: *Indurated tumor lesion with irregular borders.*

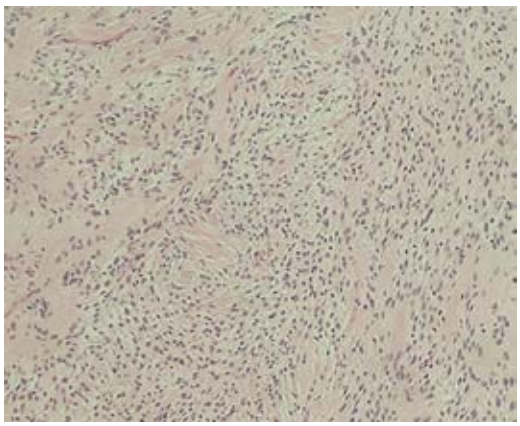

Figure 7: *Histopathologic section showing Antoni-A areas containing Verocay bodies, consisting of cells with oval-shaped nuclei and hypercellularity, and Antoni-B areas with diffuse laxity with hyaline degeneration and hypocellularity.*

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