SCHWANNOMA IN CERVICOTHORACIC TOPOGRAPHY: AN UNUSUAL CASE REPORT

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Recebido em 06/02/2020. **Aceito** para publicação em 03/03/2020

RESUMO

O Schwannoma cervicotorácico é uma neoplasia neurogênica benigna, a qual surge a partir das células de Schwann da bainha de mielina de nervos fora do sistema nervoso central. A etiologia ainda não é estabelecida e o acometimento topográfico é variável. Estima-se que pelo menos um quarto dos casos ocorra na região da cabeça e do pescoço. Embora seja mais comum por volta da terceira e quarta décadas de vida, o relato de caso em estudo apresenta uma paciente de 17 anos, cuja sintomatologia não foi manifestada, apesar do tamanho considerável da lesão. Tais fatos evidenciam a atipia e relevância do caso clínico relatado. Esse artigo, portanto, tem por objetivo destacar a relevância do Schwannoma como diagnóstico diferencial de massas cervicais, mesmo em pacientes jovens e assintomáticos. Estima-se que um quarto dos casos ocorra na região da cabeça e do pescoço.

PALAVRAS-CHAVE: Schwannoma, Cervicotorácico; cirurgia.

ABSTRACT

Cervicothoracic schwannoma is a benign neurogenic neoplasm, which arises from the Schwann cells of the myelin sheath of nerves outside the central nervous system. The etiology is not yet established and the topographical involvement is variable. It is estimated that at least a quarter of cases occur in the region of the head and neck. Although it is more common around the third and fourth decades of life, the case report in the present study presents a 17-year-old patient, whose symptomatology has not manifested, despite the considerable size of the lesion. Such facts show how atypical and relevant the reported clinical case is. This article, therefore, aims to highlight the relevance of schwannoma as a differential diagnosis of cervical masses, even in young and asymptomatic patients.

KEYWORDS: Schwannoma, Cervicothoracic, surgery.

1. INTRODUCTION

Schwannoma, also known as neurinoma, is a neurogenic neoplasm which originates from the Schwann cells of the sheath of localized nerves outside the central nervous system. Despite the existence of several hypotheses, the etiology of the disease is still unknown and can manifest in any nerve constituted of schwann cells. In this way, several areas of the body can be affected, such as the head and the neck (responsible for 25 to 45% of the cases), mediastinum, retroperitoneum and pelvis, among others. It is characterized by a benign tumor as well as circumscribed and non-invasive disease, in which age range is variable, but more common in the third and fourth decade of life. Its surgical excision should be performed to prevent the progression and possibility of malignization[1-3].

This paper aims to report the case of schwannoma in a patient of age 17, asymptomatic, detected by the appearance of cervical mass in topography of thyroid gland.

2. CASE PRESENTATION

A 17-year-old female patient, asymptomatic. Sought care due to the appearance of cervical mass in the left paratracheal region, in the topography of the thyroid gland. It was behind the sternocleidomastoid muscle and extended to the upper mediastinum. The dough had three months of evolution, it was painless to palpation, immotile to swallowing and hardened. The patient had no neurological deficits or hoarseness.

An ultrasound examination was performed, showing a regular bordered solid nodule which was hypoechoic between regions VI and VII and compatible with proliferative (lymphoma) or thymoma diseases. A fine needle aspiration biopsy (FNAB) was performed and suggested it was a fusocelular neoplasm with morphological characteristic of benignity suggestive of schwannoma.

A computerized tomography scan revealed expansive lesion, hypothense and with precise limits located posteriorly to the left lobe of the thyroid. The injury caused tracheal deviation to the right, culminating in reduction of diameter. The observed tumor presented as an insinuated mass to the superior mediastinum onto the brachiocephalic vein and caused anterior carotid deviation of the internal jugular vein and in the other vessels, posterolateral deviation without signs of involvement. The measures of the tumor were 8.7cm x

5.8cm x 4.9cm (*figure 1*).

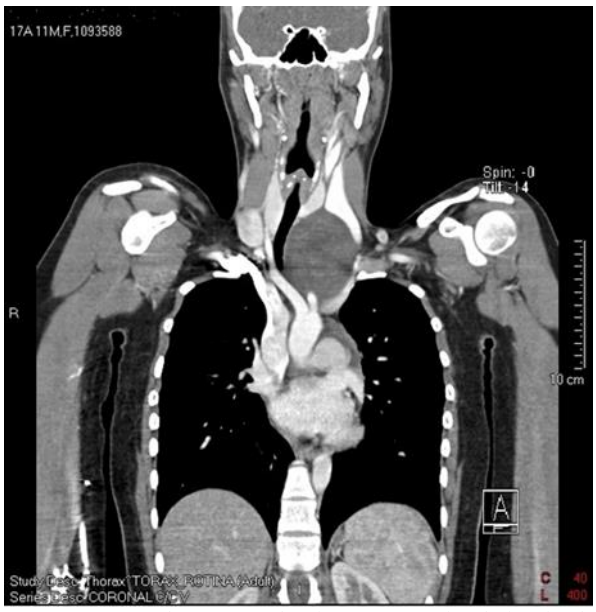

Figure 1. Coronal CT scan showing an expansive lesion in the posterior inferior cervical region of the left lobe of the thyroid with extension to the superior mediastinum.

After the diagnosis, the patient was hospitalized and submitted to surgery for the exeresis of the schwannoma. It was observed that the lesion was not invading structures and was not macroscopically associated with any nerve (Figure 2).

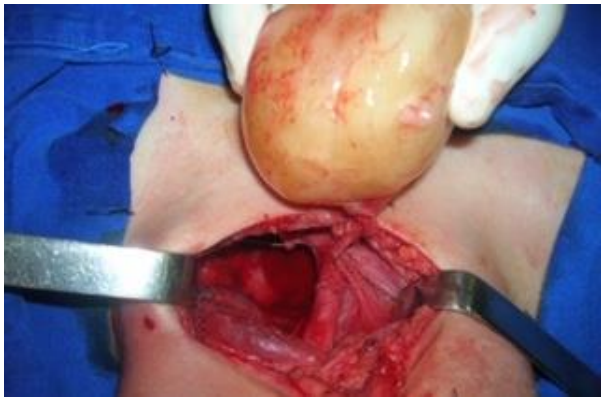

Figure 2. Operative period of tumor resection, showing tumor and location of the lesion that extended to the upper mediastinum.

During the procedure, it was found that there was total integrity of the vagus nerve, phrenic trunk, and the brachial plexus, indicating sequelae arising from the tumor. The whole procedure went smoothly and the duration was of one hour and fifteen minutes. There were no postoperative issues and the patient was discharged one day after the operation. The tumor mass removed was referred for histopathological examination. The result confirmed the diagnosis of schwannoma. The proliferation of spindle and elongated cells arranged in multidirectional bundles was witnessed, exhibiting densely cellular areas (Antoni A) and loose areas (Antoni B). In addition to that some areas with a palisade nuclear (formation of Verocay bodies) and thickened walls were observed. There were no signs of malignancy.

3. DISCUSSÃO

Schawannomas arise from the proliferation of Schwann cells, cranial nerves (except optic and olfactory nerves, which have no such cells) as well as sympathetic and peripheral nerves. They are, generally, encapsulated tumors, with the exception of those that affect the nasopharynx and the sinus. They were initially described by Verocay in 1908 and their pathogenesis not yet well understood.[4] This was only proposed in 1935, in which it was established its relation to the sheath of nerve fibers[1,2,5].

Among pairs of cranial nerves, schwannomas most commonly arise from the glossopharyngeal, accessory and hypoglossal nerves, although the involvement of the triglyceral, trochlear, facial and vagus nerves is relevant[6]. Although the etiology is still unknown some hypotheses of causal behavior of the schwannoma were raised, such as external nerve damage, spontaneous growth and radiation exposure. Approximately 25 to 45% of the schwannomas affect the head and neck region, affecting mainly a lateral region of the Neck. However they consist of less than 1% of the tumors found in this topography[1-5].

The probability of shwannoma becoming malignant ranges from 8 to 10%.[4] Although occurring in any age group, it is more common in the third and fourth decade of life[4]. Due to its slow growth and long period of quiescence, alike the case report under study, these tumors are often found at random. The most common signs and symptoms, depending on of the archaeological site involved, are pain, cough, dyspnea, weight loss, weakness, fever, hypertension, vertigo and dysphagia. However, some patients may be asymptomatic, as in the case reported, despite the considerable size of the lesion[1-3].

Commonly, these swellings are solitary and do not present a predisposition to genre. There are relations linking this tumor to schwannomatosis and neurofibromatosis. An chromosome 22 anomaly is evident in about 50% of cases, which happens in neurofibromatosis[5]. Nasosinusal schwannoma corresponds to 4% of the schwannomas of the head and neck and due to its slow growth, remains asymptomatic for a long period. When there is symptomatic manifestation, a nasal obstruction is the most frequent symptom, with pain, bone dehiscence (including the skull base) and other neural symptoms manifest themselves with growth of the tumor. However, if associated with neurofibromatosis, there may still come to be the appearance of multiple nodules, although it is still a rarely reported manifestation[4,5].

Macroscopically, the schwannomas present as a mass involved by a capsule with the diameter of less than 1 millimeter up to 7 centimeters[7]. The present report, however, surpasses that commonly described in literature. These tumors may have cystic areas and parenchymal consistency hardened non-infiltrative character. Histologically, three different tissue patterns: Antoni A and Antoni B, or a mixture of the two subtypes. The Antoni A pattern is more typical in schwannomas

and it is characterized by highly cellular regions with the presence of elongated spindle cells, which can be found in a well-ordered and dense setting, in which they are arranged in single palisade nuclei originating the Bodies of Verocay. The Antoni B fabric has degenerative and mixidematose, containing a lower cellularity, arranged in an irregular setting and the occurrence of microcysts in some cases[8-10].

Considering the morphological aspects, the differential diagnosis should also contemplate other soft tissue tumors, such as fibroma, chondroma, and leiomyoma[11]. Cytological examination obtained by FNAB may be suggested. If this is not conclusive, the next procedure is to remove the nodule for evaluation by freezing and/or conventional anatomopathological examination, in order to determine the malignancy or benignity of the tumor. The technique of immunohistochemistry can be applied by observing the existence of the S-100 protein, which is found only in Schwann cells and melanocytes localized in soft tissues outside the central nervous system, being absent in malignant neurinomas. The S-100 protein evidences neuroectodermal origin of the tumor[5-7].

Treatment consists of complete surgical excision of the tumor. As the schwannoma is not radiosensitive, the possibility of radiotherapy is ruled out. In the cases of upper airway schwannomas, depending on the extent of neoplastic mass, the surgical approach can be done by endoscopy, which has safe procedure for small tumors. However, this technique does not apply to multiple tumors that extend into the deeper layers of the tissue. In cases where surgical access is external, attention must be paid to the risk of injury to the recurrent laryngeal nerve. Also, due to the proximity of the fibers of the vagus nerve in this region, complete surgical excision is a challenge[7,9,11,12].

The patient should be advised about possible postoperative neurological sequelae, such as Horner's syndrome and vocal paralysis, among others. Despite being uncommon, there are also reports of Harlequin syndrome, arising from cervical sympathetic nerves and concomitant damage of the upper cervical ganglion during the surgical procedure for cervical Schwannoma excision. Tumor recidivism is low, and there is a significant association with the degree of mitotic counting. Slow tumor growth, very low recurrence rate and non-invasive nature indicate a good prognosis[1,7,13-15].

4. CONCLUSION

Although not very common, the possibility of cervical schwannoma should be considered in cases of changes in utterance and deglutition, coughing and onset of mass in the neck region. Differential diagnosis is important to exclude hypothesis of other neoplasms due to the not very specific morphological aspect of the neurinomas, which can be easily elucidated during histological analysis. The early exertion of the lesion and the use of adequate technique are determining factors for good prognosis, contributing to better life quality to the patient. In a nutshell, the age of onset and time of tumor progression in the presented report justify the relevance of the case.

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