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Case Report

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Cervical Thymoma

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ABSTRACT

Context: Cervical thymoma is a primitive thymic neoplasia. It is very rare. This disease presents higher incidence in female patients in their 4th to 6th decade of life. We present a case report of a cervical thymoma

Case Report: 54-year-old female patient, caucasian, with no history of morbidity, presenting a left cervical nodule close to the thyroid gland. During the 30 months of investigation a left cervical nodule grew progressively next to the thyroid while the patient showed no symptoms, making accurate diagnosis difficult. Tests on her thyroid function did not show changes, nor were there changes in any subsidiary tests. The diagnosis of the disease was made intra-operatively through total thyroid individualization. The results were confirmed by the histological findings from the ressected material. Cervical thymoma is a very rare disease, with difficult preoperatory diagnosis. Some additional study methods which are employed today are thallium 201, technetium 99 and iodine 131 scintigraphy, magnetic nuclear resonance and especially histopathological findings and classification.

Uniterms: Neck. Cervical thymoma.

INTRODUCTION

Cervical thymoma is a primitive thymic neoplasia. It is very rare, with only 18 cases having been reported up to 1994 according to a survey of English-language literature, of which 16 cases were female.[1] This disease presents higher incidence in female patients in their 4th to 6th decade of life.[2]

The thymus is a gland originating from the epithelium, which forms in the third portion of the pharynx during the tissue matching that takes place in the sixth week of fetal life. In the eighth week of fetal life there is a migration of the thymus from the cervical region towards the caudal and medial region to its final location at the upper mediastinum. Failure of this migration will cause abnormalities of thymic tissue such as hypoplasia, cysts, glandular tissue at the periphery of the thyroid and cervical thymus with or without mediastinal extension.

Thymic tissue with atypical location has been classified[3] into seven types: type 0, with no cervical extension; type 1, right or left extension, without contact with the thyroid gland; type 2, left or right extension in contact with thyroid; type 3, bilateral extension without contact with thyroid; type 4, bilateral extension, one side in contact with thyroid; type 5, bilateral

extension in contact with thyroid; type 6, unilateral extension, above the thyroid gland; type 7, discreet nodes at lateral positions in contact with the thyroid. Types 3 and 4 are the most frequent presentations. The case reported here is classified as type 2 thymic tissue.

CASE REPORT

The patient was female, 54 years old, Caucasian, with no history of morbidity, presenting a left cervical nodule close to the thyroid gland. On February 24, 1994, a thyroid scintigram was made that showed a normal thyroid gland and an extra thyroid nodule, which was diagnosed as benign and was referred for conservative treatment at another institution. After two and a half years of nodule growth, another thyroid scintigram was produced and the report showed an enlarged thyroid gland (nodular goiter) and a "cold" cervical nodule, as it was not possible to ascertain whether this was a thyroid nodule by scintigraphy.

During its evolution the patient was asymptomatic and did not show any cervical masses other than the one reported above. Upon examination of the nodule, the thyroid tests were normal and chest x-rays showed no changes.

On July 21, 1997, she was submitted to surgical treatment with total resection of the cervical mass in contact with the upper mediastinum. After performing a necklace-type incision 3 cm above the sternal furcula, the mass was observed in the upper mediastinum, projected towards the suprasternal and pre-thyroid regions. We had to perform blunt detachment techniques in order to free the tumor, which presented a solid aspect with a whitish brown color. The postoperative period was uneventful.

Macroscopic examination

The surgical sample was nodular, measuring 6.0 cm x 5.0 cm x 4.0 cm, lobulate, whitish brown color, hard, woody, covered by a delicate membrane. At cutting, we observed a fasciculated surface, milky color with whitish stria (Figure 1).

Microscopic examination

The cuts showed epithelial and lymphocytic elements uniformly spread, with alternate predominance. The existence of a tumor surrounded by a compact connective tissue capsule, interspersed by numerous partially hyalinized connective septa was confirmed. Epithelial cells presented clear cytoplasm, vesiculate ovoid nuclei grouped in small numbers of cells, or were seen with spaces amid the lymphocytes. Some of these cells had mitotic activity. However, we did not observe any atypical cellular element in the material examined (Figures 2 and 3).

DISCUSSION

Thymic neoplasia takes place predominantly in the anterior mediastine and is usually a tumor

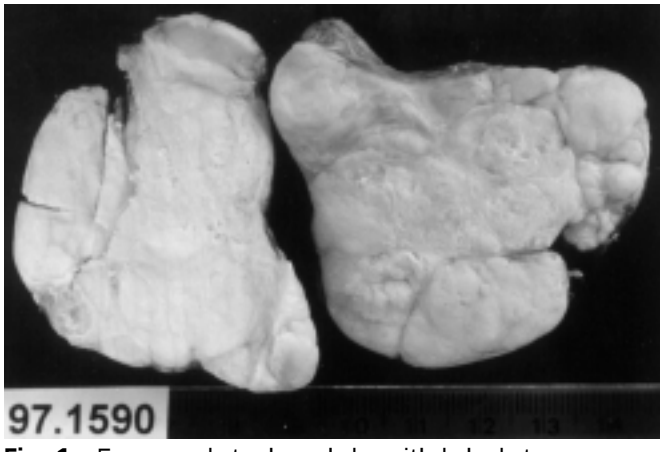

Fig. 1 - Encapsulated nodule with lobulate appearance.

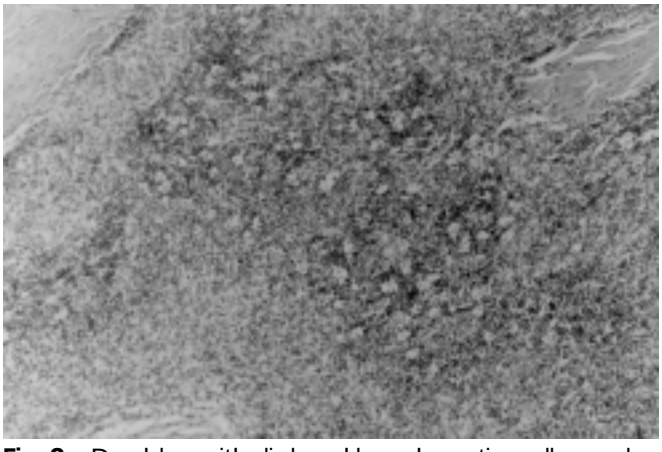

Fig. 2 - Double epithelial and lymphocytic cell population, with "starry sky" appearance (Hematoxylin Eosine).

with a slow growth rate.[4,5] It occurs in around 50% of the patients with miastenia gravis and is more prevalent in female patients.[2]

The majority of cervical thymomas present asymptomatic evolution, becoming painful in the cases where there is local infection or very rapid growth together with dysphagia. Cervical thymic tissue may be found in about 21 to 42% of children and occasionally it mimics a cervical tumor.[1] There is no report of an incidence ratio between men and women with this disease due to its low incidence in male patients.

The case which we present in this paper confirms the clinical status reported in world literature, with asymptomatic history, slow growth and diagnosis confirmed by pathology. The differential diagnosis between thyroid nodule or cervical tumor was made at the intraoperatory evaluation from the position of the tumor, its total individualization from the thyroid, its macroscopic aspect and its relationship with the upper mediastinum. The diagnosis was confirmed via anatomopathological studies.

The largest study to date was carried out in 1983 by Yamashita et al[6] with 12 cases of cervical thymomas in a group of 657 patients with Basedow's disease, by performing histopathological and epidemiological evaluation of this pathological entity.

Investigation techniques for cervical nodules have needed to be altered, especially when tests have been inconclusive. Vangrove et al[1] reported a case in which the diagnosis of cervical mass could only be decided on by using histopathological study of the aspiration biopsy with use of immunoperoxidase, whereas the initial evaluation had suggested lymphocytic chronic thyroiditis or malignant lymphoma. Nomori et al[7] reported on the heterogeneity of the complicated histology in thymic neoplasia for diagnostic purposes and specific and precise treatment.

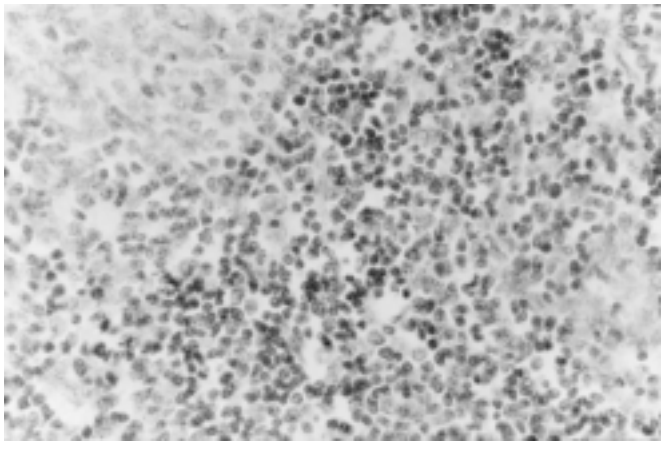

Fig. 3 - Detail of the double cell population, with epithelial cells in evidence (Hematoxylin Eosine).

Scintigraphy with thallium 201 is used for the differential diagnosis between thymoma and thyroid neoplasia, according to Fukuda.[8] Technetium 99 is used similarly according to Miller.[9] We used radioactive iodine with our patients.

According to Kiyosue et al,[3] magnetic nuclear resonance should be the examination of choice for investigating cervical masses or nodules, especially in cases where there is communication among such entities and the mediastinum, raising the suspicion of a cervical thymoma.

According to the study performed by Ho et al,[2] the histogenetic classification of thymic epithelial tumors is directly related to the patient's survival rate. Five groups have been reported: group 1: medullar thymoma and mixed thymoma (100% survival over 10 years); group 2: predominantly cortical thymoma (100% survival over 10 years); group 3: cortical thymoma and well differentiated thymic carcinoma (40% survival over 10 years); group 4: other thymic carcinomas (30% survival over 10 years); group 5: unclasssifiable. The pathological classification has predictive value in evaluating the aggresiveness potential of these tumors[4] and therefore the prognosis for these patients, thus contributing to appropriate post-operative treatment. Our patient was histogenically classified as being group 1.

Of the different histopathologic classifications for thymomas, none is totally satisfactory. Several studies have found no correlation between histological type and prognosis, while other studies clearly mention this correlation.

As cervical thymoma is a rare disease with pre-surgical diagnostic difficulties, immunohisto-chemistry provides a more efficient method for diagnostic investigation and adequate intervention

in the treatment of patients with this disease. At present there are no specific markers for cervical thymomas, but studies investigating potential markers are promising and in the future these may become part of the routine tests in pathological investigation of such tumors.

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RESUMO

Contexto: O timoma cervical é neoplasia primitiva do timo. Sua incidência é muito rara. Esta doença apresenta maior incidência em pacientes do sexo feminino, entre a quarta e sexta décadas. **Relato de Caso:** Apresentamos o caso de uma paciente do sexo feminino, 54 anos, com timoma cervical. Durante o período de dois anos e meio de investigação teve evolução assintomática com crescimento progressivo do nódulo cervical, próximo à tireóide. A paciente não apresentou alterações dos exames de função tireoidiana ou dos exames subsidiários realizados. O diagnóstico de timoma cervical foi realizado no intra-operatório sendo conclusivo com o estudo anatomopatológico da peça retirada. O timoma cervical é raro, com difícil diagnóstico pré-operatório. Atualmente utiliza-se métodos complementares para o diagnóstico preciso como: a cintilografia com Tálio 201, o Tecnésio 99 e o Iodo 131, além da ressonância nuclear magnética e, principalmente, realizando o estudo e classificação histopatológica.