Solitary fibrous tumor of the thyroid. Report of a case with unusual clinical and morphological findings

ABSTRACT

The Solitary fibrous tumor (SFT) of the Thyroid, is an unusual slow-growing mesenchymal tumor mostly found during adulthood, that exhibits a favorable biological behavior. This case of SFT of the thyroid describe the clinical and immunohistochemical analysis that will provide a closer interpretation of this pathology, regardless its rarity. A rapidly growing cervical mass that ascends with swallowing is presented. Ultrasound classification system TI-RADS-4 is determined, and Fine Needle Aspiration (FNA) is performed, morphological studies presented a distinctive proliferation of cells with an epithelial habit without a defined pattern. Expression markers such as STAT-6, BCL-2 and CD34 are essential for the diagnosis, along with the negativity of others, and molecular confirmation with NAB2-STAT6. Solitary fibrous tumor (SFT) of the Thyroid with these morphological characteristics are poorly described in the literature representing a challenging diagnosis.

Keywords: SFT, Thyroid, Solitary Fibrous Tumor, immunohistochemical, Case Report.

Caso clínico

1. Introduction

Solitary fibrous tumor (SFT) of the thyroid is a mesenchymal neoplasm of fibroblastic lineage of unknown etiology that shows a morphology characterized by spindle cells with variable stroma and an ectatic vascular network reminiscent of the hemangiopericytic-like pattern (1). It was first described in the pleura by Wagner in 1870 (2), however it can originate in any location (soft tissues, organs of the thoracic/abdominal cavity, pelvic cavity, bone tissue as well as in various locations of the head and neck (3).

The Solitary fibrous tumor (SFT) of the thyroid was first described in 1993 by Tacagni (4). It is an exceptional location, where only less than 40 cases have been described in the literature, <0.1% arise in the head and neck (5, 6). According to epidemiological data, it can occur in both sexes with a variable age range between 28 to 80 years with no known risk factor. Clinically it presents as a slow-growing painless lesion, however, on rare occasions, it can show hypoglycemia as part of a paraneoplastic syndrome (Doege-Potter syndrome) in less than 10% of published cases (7). 90% of these lesions are benign, with a very low rate of recurrence and metastasis, and the remaining 10% show malignant potential (6).

The morphological characteristics are well established. They are spindle-shaped or fibroblast-like cells with variable stroma, which can adopt different architecture. The "Patternless Pattern" and the hemangiopericytic-like pattern are the most frequent, but there are others such as whorled, wavy, desmoid and palisade with little mitotic activity (8). The characteristic immunohistochemical study of this lesion is the expression of STAT-6, CD34 and BCL-2 together with the negativity of other markers (8). Currently, a genetic fusion characteristic of this neoplasia is recognized, the fusion of the NAB2-STAT6 genes (7, 8).

2. Methods

We present this patient's case, conducted at The Doctor Peset Hospital, Valencia - Spain in June 2021. The Case report was approved by the Dr. Peset Hospital Chancellery of universal health, Public Health of Valencia Spain (Documented Reference on 06/21) history, clinical findings, immunohistochemical analysis, cytopathological data, management, and resolution has been also approved by a signed informed consent. In addition to the case, we execute a review of the previously published cases of STF and consolidate terms and knowledge.

3. Results

A 46-year-old male with no relevant clinical history, presented with a painless right latero-cervical mass, 20 days old, with no other associated symptoms. On physical examination, the lesion is not adherent to deep layers and ascends with swallowing, without palpation of lymph nodes. Thyroid hormones are within normal parameters. In the imaging study, the ultrasound confirmed a solid 4 x 4 cm lesion in the right thyroid lobe, classifying it according to the TI-RADS 4 thyroid ultrasound classification system, for which an echo fine needle aspiration (FNA) was performed. The cytological diagnosis is of a poorly differentiated tumor and, subsequently a histological study is performed after excision of the lesion.

3.1 Macroscopic study

Right hemi-thyroid weighing 42 g and measuring 6 x 4 x 4 cm. On section, a homogeneous, pseudo-encapsulated whitish lesion is identified, which occupies almost the entire thyroid lobe (Figure 1).

3.2 Microscopic study

Four months after the histological diagnosis, PET/CT is performed and a right paravertebral hypermetabolic lesion is evident, extending from the base of the skull to C3, and multiple liver lesions, which it is biopsied to determine their origin (Figure 2).

Figure 1. Gross. Gross Figure of a cut section of thyroid gland, showing a solid grayish-white nodule that replaces almost the entire glandular parenchyma.

Palabras clave: Tiroides; Tumor Fibroso Solitario; inmunohistoquímica; Reporte de un caso.
Four months after the histological diagnosis, PET/CT is performed and a right paravertebral hypermetabolic lesion is evident, extending from the base of the skull to C3, and multiple liver lesions, which it is biopsied to determine their origin. (Figures 3, 4), (Table 1).

**Figure 2.** Morphology and IHC of the thyroid lesion. (A-B-C-D-E-F-G-H)
A: Loss of thyroid architecture due to the presence of a tumor with a solid pattern.
B: Cells with epithelial habit.
C: Cells with extensive eosinophilic cytoplasm and hypochromatic nucleus.
D: Irregular vascular structures.
E: Thyroglobulin.
F: PAX-8.
Figure 3. Morphology of the skin lesion, (I–J–K–L).
I: Well-defined tumor that occupies the dermis and extends to the hypodermis.
J: Proliferation of spindle cells and other epithelial cells with focal atypia.
K: Occasional vascular structures of irregular morphology without nuclear atypicality.
L: Hypocellular/hypercellular areas with hyaline stroma.

Figure 4. Morphology and IHC of the liver lesion, (M–N–O–P).
M: Spindle cell proliferation.
N: Spindle cells arranged in short fascicles with focal nuclear polymorphism and mitosis.
O: CD34 +.
P: Ki67 high.
Table 1. Morphological characteristics of soft tissue and liver biopsies.

<table>
<thead>
<tr>
<th>CHARACTERISTIC</th>
<th>SOFT PART BIOPSY</th>
<th>LIVER BIOPSY</th>
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<tbody>
<tr>
<td>Morphology</td>
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<td>Pattern</td>
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</tr>
<tr>
<td>IHC</td>
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<td></td>
<td>CD99+</td>
<td>CD99+</td>
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<tr>
<td></td>
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<td>Ki-67 30%</td>
<td>Ki-67 80%</td>
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The three lesions share expression of CD34, CD99, STAT-6 and negativity for CKs, TTF-1, thyroglobulin, PAX-8, calcitonin, S-100, melan-A, HMB-45, actin, desmin, DOG-1, CD117, CD31 and ALK, as well as expression of the NAB2 exon 6 /STAT6 exon 16 fusion gene (RT-PCR).

4. Discussion

Thyroid Solitary fibrous tumor (SFT), is characterized by monomorphic spindle cells with soft nuclei accompanied by stroma and ectatic vascular structures reminiscent of the hemangiopericytic pattern, related to non-aggressive biological behavior. However, histological criteria associated with a negative prognostic factor have been described, such as hypercellularity, >4 mitoses, necrosis, nuclear atypia, and cell dedifferentiation. In our case, the thyroid lesion showed a poorly differentiated epithelial cell proliferation, without necrosis or >3 mitoses x 10 AGC. It caught our attention that four months after diagnosis, soft tissue and liver lesions present with atypical spindle cell morphology, with a hemangiopericytic pattern and a considerable increase in mitoses compared to the thyroid lesion. In the literature, there are few cases of thyroid SFT that meet these characteristics, one of them with recurrence and metastatic disease at 5 months of follow-up (9, 10, 11, 12).

Among the differential diagnoses are primary thyroid carcinomas (medullary carcinoma, anaplastic carcinoma, and poorly differentiated thyroid carcinoma). Medullary carcinoma shows a wide morphologic variety and expresses calcitonin, chromogranin, and synaptophysin; anaplastic carcinoma is an aggressive, fast-growing tumor that presents clearly malignant characteristics, a high rate of mitoses and great pleomorphism, and expresses epithelial markers, being CD34 negative; and poorly differentiated thyroid carcinoma, characterized by nests of monomorphic cells, without pleomorphism, that express epithelial markers and are also CD34 negative.

We must also include in the differential Figure 1(3) support and diagnosis of some mesenchymal tumors such as: monophasic synovial sarcoma, leiomyoma, schwannoma/neurofibroma and malignant peripheral nerve sheath tumor. Monophasic synovial sarcoma is morphologically like Solitary fibrous tumor (SFT) that expresses BCL-2 and CD99, but CD34 is negative; the leiomyoma expresses muscle markers such as desmin and actin; schwannomas/neurofibromas are usually spindle cell neoplasms with an undulating pattern, the first with Antoni A and Antoni B areas and Verocay bodies and the second with mixed cellularity, both S-100 positive and CD34 negative; and malignant peripheral nerve sheath tumor showing high-grade morphology (11, 12).

5. Conclusions

Solitary fibrous tumor (SFT) of the Thyroid, is an exceptional indolent entity, clinical follow-up being essential for a rare but possible recurrence if not excised and emphasizing on the morphological details that are associated with aggressive behavior to give a proper management and its rare metastasis prevention.

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Conflict of Interest

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References


