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Myxoid and Lipomatous Solitary Fibrous Tumor of Soft Tissue: A Case Description

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Abstract

We present a case of a myxoid and lipomatous solitary fibrous tumor that was observed in a 67-year-old man. The tumor, which had a maximum diameter of 10 cm, was located in the soft tissues of the dorsal region and appeared macroscopically well delimited and encapsulated. Upon cutting, a markedly gelatinous internal surface was observed. A microscopic study revealed an intense and diffusely myxoid neoplasia, with small areas of adipose aspect, in which histological (staghorn vessels, perivascular hyalinization, fusiform cells of benign aspect) and immunohistochemical (intensive positivity for CD34, Bcl-2 and Cd99 and negativity for muscle markers) data were consistent with a solitary fibrous tumor were observed. To conclude, the main characteristics of this lesion are discussed, and a differential diagnosis is established with other entities.

Keywords

Solitary Fibrous Tumor, Myxoid and Lipomatous Differentiation, Histopathology, Immunohistochemistry

1. Introduction

The constant development of oncology and all of the branches of medicine involved in the treatment of cancer have led to the introduction of increasingly effective and personalized modern therapies in which exact knowledge of the tumor is of great importance. As a result, we pathologists find ourselves obliged to make increasingly accurate diagnoses, and therefore, to exhaustively determine neoplastic pathology, both of more frequent cases and of those variants are rarely observed in daily clinical practice.

Based on this affirmation, we were encouraged to present the case set forth below: A myxoid and lipomatous solitary fibrous tumor. A rare histological variant, rarely described in the medical literature, was observed in a 67-year-old man.

2. Clinical Case

A 67-year-old man, without a history of interest, came to our hospital because of a slow-growing tumor that was located in the dorsal region and had increased in size in recent months.

Radiological studies revealed the solid character of the lesion, its good delimitation and its superficial intramuscular location.

Once completely removed, an irregularly elongated tumor was observed that was apparently well delimited and encapsulated; it was of soft consistency and reached a maximum diameter of 10 cm. Upon cutting, its marked gelatinous aspect was evident (**Figure 1**).

A histological study showed an markedly myxoid neoplasia, of low cell density, in which the presence of abundant elongated and branched vessels (staghorn vessels) were noted. Neoplastic cells were of a homogenous and fusiform appearance; benignity was suggested because mitosis was not observed. After extensive sampling, small occasional pockets of adipose cells were observed (**Figure 2**).

An immunohistochemical study demonstrated intense and diffuse positivity of the cancer cells for CD34, CD99, Bcl-2, EMA and vimentin; moreover, it was negative for actin, desmin, H-caldesmon and MyOD1. S100 staining showed only positivity in adipose cells. The proliferative activity (MIB-I) was lower than 1% (**Figure 3**).

Based on these findings, our diagnosis was of myxoid and lipomatous solitary fibrous tumor (hemangiopericytoma).

Two years after its removal, no signs of recurrence or metastasis are observed

3. Discussion

Since its first description at the pleural level [1], solitary fibrous tumor has been a constantly expanding entity, both because it has been observed in multiple extrapleural

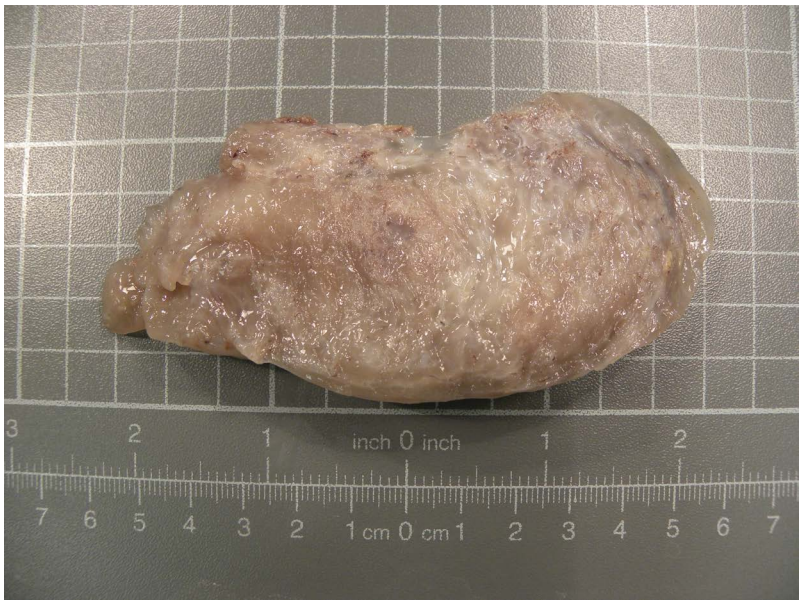
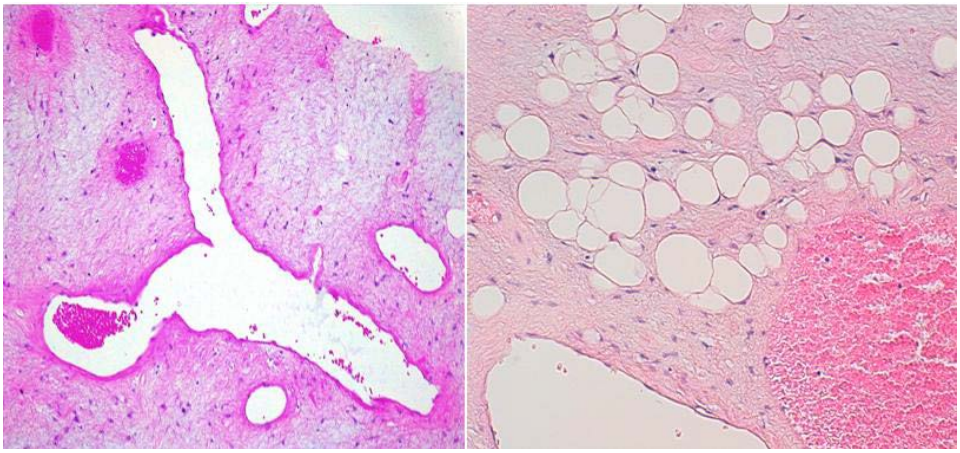
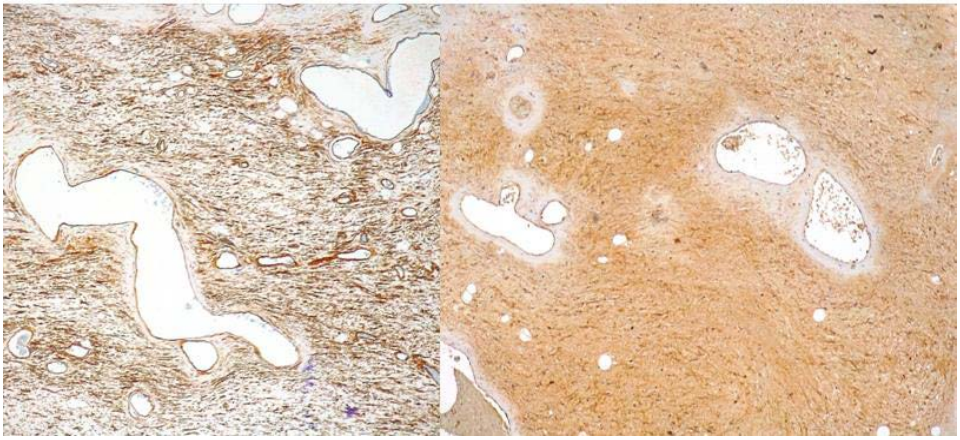


Figure 1. Myxoid and lipomatous SFT (Macros): Note the good delineation of the tumor and its gelatinous appearance.



(a) (b)

Figure 2. Myxoid and lipomatous SFT: Neoplasia markedly myxoid with staghorn vessels (a) and occasional fat cells (b). ((a) HE×100), ((b) HE×400).



(a) (b)

Figure 3. Myxoid and lipomatous SFT: Positivity of tumor cells for CD34 (a) and CD99 (b) ((a) and (b) IH×50).

locations [2] [3] and because of its ability to include lesions that traditionally had received other denominations. Examples of the latter include angiofibroma of giant cells, which is a neoplasm that is generally located in the orbital region [4], and hemangiopericytoma, or its variant lipomatous hemangiopericytoma, which is primarily located in soft tissues [5] [6]. A thorough review of these entities, based on the study of their clinical behavior and ultrastructural and immunohistochemical properties, have led most authors to consider them as variants of the solitary fibrous tumor; thus, its old nomenclature has become obsolete [7] [8].

In our case, we are facing a soft tissue tumor with typical hemangiopericytomatous vascularization (staghorn vessels) and cellularity of benign appearance, whose immunohistochemical profile (CD34+, CD99+, Bcl-2+, EMA+, actin−) is completely consistent with a solitary fibrous tumor [9]. In our view, the most striking feature of this tumor was its marked and diffuse myxoid character and, to a lesser degree, its focal adipose differentiation.

This myxoid change, which can be observed locally in many cases, takes on exceptional character when it is as intense as the tumor in this study [10] and is also accompanied by areas of lipomatous aspect. This appearance can cause diagnostic uncertainty, requiring the pathologist to make a differential diagnosis with other entities [11] [12]. Thus, in our example, it was necessary, together with extensive sampling of the piece to confirm the myxoid character of the lesion over its entire surface, to establish a differential diagnosis with entities such as intramuscular myxoma (absence of prominent vessels, CD34−), angiomyxolipoma (larger amount of adipose tissue, CD34−), myxoid liposarcoma (larger proportion of adipose tissue, presence of lipoblasts, CD34−), myxofibrosarcoma (histological aggressiveness, CD34−, absence of prominent vessels), muscular tumors (leiomyoma and leiomyosarcoma) with myxoid differentiation (CD34−, actin+, desmin+), nerve tumors (neurofibroma, nerve sheath myxoma) that are positive for S100, pleomorphic hyalinizing angiectactic tumor (pleomorphism, CD99−, Bcl-2−) or myxoid chondrosarcoma (CD34−, absence of prominent vessels, cellularity in rows).

Molecular analyses have discovered that almost all SFTs harbor an NAB2-STAT6 fusion gene, which is considered specific to this tumor type and recent

Studies have suggested that nuclear STAT6 immunoreactivity is a highly sensitive and specific marker of SFTs and can be helpful when diagnosis is inconclusive by conventional methods [13].

Once the nature of the neoplasm was established (solitary fibrous tumor), both its macroscopic appearance (good delimitation) and microscopic features (absence of necrosis and mitosis, low proliferative index and absence of cytologic atypia) were indicative elements of a favorable prognosis.

The clinical evolution confirms, for the moment, the histological findings. Two years after its removal, no signs of recurrence or metastasis are observed and the patient shows a good general condition, Behavior similar to that described by other authors, in cases histologically similar to ours.

4. Conclusion

Finally, we briefly emphasize the importance both for clinicians and pathologists to study, stay up to date and continue their ongoing collaboration because these are key elements for proper patient care.

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