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Superficial CD34-Positive Fibroblastic Tumor: First Case Report from Latin-America and Review of the Literature

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Abstract

Superficial CD34-positive fibroblastic tumor is an infrequent soft-tissue tumor characterized by a lesion located in the subcutaneous fascia without affecting the soft or deep muscle planes. Its incidence is unknown; therefore, it represents a diagnostic challenge for both the surgical oncologist and pathologist. Given its high risk of local recurrence, extensive surgical resection is the recommended treatment modality. This is the first report of this rare entity in Latin-America.

Keywords: CD34; fibroblastic tumor; immunohistochemistry; diagnosis; soft tissue neoplasm

Introduction

The superficial CD34-positive fibroblastic tumor is a rare soft tissue lesion [1,2], that primarily affects the subcutaneous fascia without involving deep muscle tissues [3,4]. It predominantly occurs in the lower extremities of adults. However, due to its rarity, the current incidence rate of this tumor remains unknown. Currently, considering its risk of local recurrence, the most appropriate treatment has been extensive tumor resection. [3,5]. This review aims to provide an overview of the diagnostic challenges associated with this lesion and suggest potential improvements in its management.

Case Presentation

A 29-year-old male patient who presented to the Clinica Universitaria Colombia in Bogota-Colombia with one-year history of pain in the right axillary region associated with a progressively larger mass extended to the pectoral region. The mass had a hard, rubbery consistency, tender to palpation, a red punctate lesion on the skin surface and measuring 4 cm of diameter. Soft tissue ultrasound showed changes suggesting a subcutaneous cystic lesion in the axillary tail of the right breast and inflammatory looking right axillary lymphadenopathy.

The patient underwent excisional biopsy, intraoperatively there was a 7x4 cm lesion, without muscular or vascular involvement. The initial pathology analysis showed atypical neoplasia with atypical fibrohistiocytic characteristics and prominent inflammation (Figure 1). A second review by an extra-institutional expert in soft tissue tumor pathology from the Brigham and Women's Hospital reported a markedly pleomorphic tumor formed by fusiform, polygonal, or multinucleated cells with irregular nuclei and variable presence of prominent nucleoli. Despite the remarkable degree of pleomorphism, mitosis was very difficult to identify, and a prominent mixed inflammatory infiltrate was found with numerous histiocytes.

Immunohistochemistry studies showed intense and diffuse CD34 reactivity of the tumor cells (Figure 2) with associated desmin reactivity, and a Ki-67 index of 20%. Additionally, there was marked lymphoid infiltrate accompanying the lesion (Figure 3). The other markers used (pancytokeratin, protein S100, SOX10, AML, P16, P53, HMB-45, EMA, and STAT6) were negative. The tumor was reported as superficial CD34-positive fibroblastic tumor.

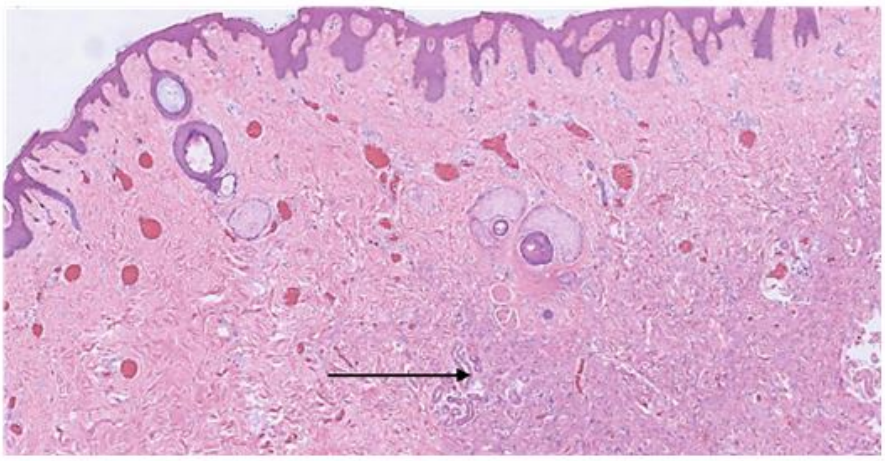

Image1: Skin with unaltered epidermis, showing a poorly circumscribed lesion in the dermis, with the presence of cells exhibiting marked pleomorphism, of fusiform appearance, irregular nuclei, evident nucleoli, and eosinophilic cytoplasm. Frequent mitotic figures are not seen, and the lesion is accompanied by ectatic capillaries and abundant lymphohistiocytic infiltrate.

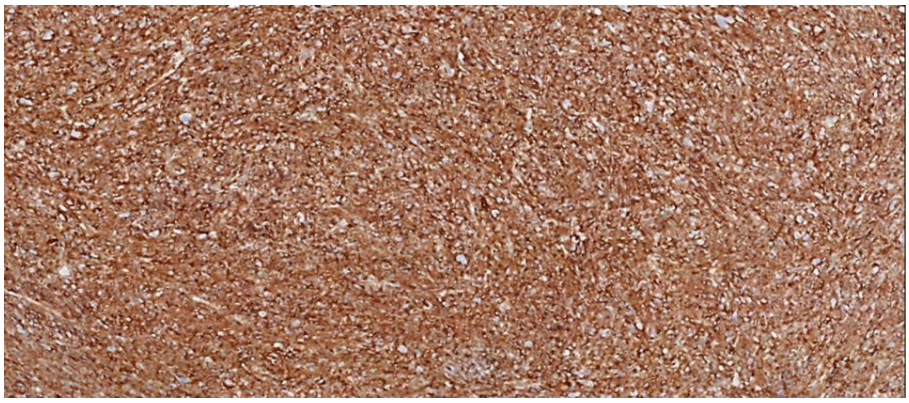

Image2: Strong diffuse reactivity of CD34 marker in immunochemistry stain.

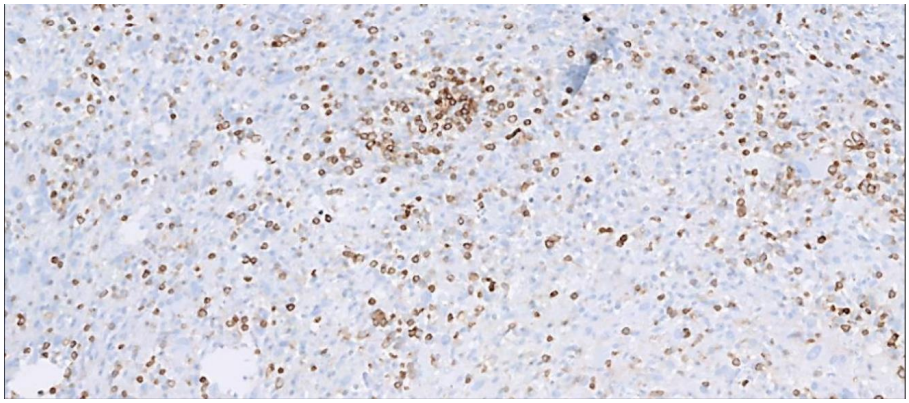

Image3: Positive CD45 marker in lymphoid infiltrate accompanying the lesion.

Based on the histological findings, additional imaging was performed with chest and abdominal computational tomography (CT), which did not show evidence of metastatic disease. Decision was made to take the patient for a repeat wide local excision with 1cm margins and ipsilateral axillary lymphadenectomy. Pathology reported negative margins. The case was then discussed in the institutional multidisciplinary tumor conference, surgical treatment was deemed sufficient without indication for adjuvant therapy. Lastly, the patient was followed for 24 months without evidence of recurrence.

Data Availability

The authors confirm that the data supporting the findings of this study are available within the article and its supplementary materials.

Discussion

CD34-positive fibroblastic tumor was first described by Carter et el. in a series of 18 cases [1]. The mass has been described as a slow growth, firm mass of yellow coloration and gelatinous appearance, with diameter ranging from 1.5 to 10 cm (mean of 4.1 cm), commonly affecting adults between 20 to 76 years (mean of 38 years) without gender preference [1,6,7].

In two thirds of the cases, tumors occurred in the lower extremities [1,4,8]. Given that only 44 cases have been reported in the literature so far, there is not clear data about its incidence on the general population. Moreover, due to the difficulty in diagnosing this type of tumor, we suspect that the incidence rate is much higher and that these tumors have been historically classified as undifferentiated pleomorphic sarcomas or myxofibrosarcomas, among other differential diagnoses with similar presentation and pathologic characteristics [4,9].

Superficial CD34-positive fibroblastic tumor is characterized by spindle and/or fusiform cells, which exhibit high pleomorphism with prominent nucleoli, frequently accompanied by xanthomatous histiocytes and mast cells. These cells show strong positivity for CD34, and some of them react weakly with pankeratin markers. On the other hand, since it is a spindle cell mesenchymal neoplasm, other vascular markers different from CD34, such as CD31 and FL1, are negative, as well as the melanocytic, myogenic, and neural markers [10,11].

In addition, findings such as significant nuclear pleomorphism, low mitosis rate <1/50, and a Ki-67 index between 1-5% can be found. The tumor often lacks expression of S-100, α-smooth muscle actin, and TP53 markers [1,6,9]. Cytogenetic studies revealed a translocation of t(2;5) (q31;q31) in tumor cells [6].

Due to its marked pleomorphism, these tumors are often misdiagnosed as other etiologies with frequent pleomorphism, such as atypical fibroxanthoma, undifferentiated pleomorphic sarcoma, epithelioid sarcoma, dermatofibrosarcoma protuberans, among others. However, due to the low Ki-67 cell proliferation index, negative myogenic, neural, or vascular lineage markers, and the low mitotic index, this neoplasm does not fit into the previously mentioned diagnoses. Therefore, it constitutes a diagnostic challenge for pathologists and surgeons.

Given that these tumors usually manifest as slow-growing superficial soft tissue masses, several years may elapse between their appearance and diagnosis. In addition, most patients undergo surgical resection based on the findings at clinical examination without any extension studies.

Mao et al. grouped and described the characteristics of the cases reported in a literature review published in 2020 [3]. The authors included 39 patients, with a male to female ratio of 1.5:1, ranging from 16 to 76 years of age, with a predominant location in the lower extremities, especially in thighs and gluteus [8]. Clinical follow-up was not reported in all patients and varied from one to 104 months with an average duration of 24 months. Within the case series described, only one patient presented with metastasis to the external iliac lymph node chain seven years after the primary tumor resection. The rate of survival was 100% without evidence of recurrence at the end of the follow-up.

In this report we presented the first case of superficial CD34-positive fibroblastic tumor treated in Latin-America. Our treatment strategy was comparable to the ones reported in the literature with wide local excision and local lymphadenectomy without evidence of tumor recurrence at 24-month follow-up.

Conclusion

Superficial CD34-positive fibroblastic tumor is a rare soft tissue lesion that presents diagnostic challenges. Its unique histological features and clinical behavior necessitate careful evaluation and multidisciplinary management. Establishing an international registry, conducting genetic and molecular studies, promoting collaboration, and conducting long-term follow-up studies are essential for advancing our understanding of this rare tumor and optimizing patient care.

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