CASE REPORT

Poorly differentiated synovial sarcoma in the wrist - Case report*

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Abstract: Synovial sarcomas are rare malignant tumors affecting mainly young adults, presenting as a slow growth mass located in deep soft tissues of extremities, near the joints. In this report a 34-year-old male patient, presented an ulcerovegetative lesion on the right wrist which was completely excised. Histopathology and immunohistochemistry confirmed synovial sarcomas with poorly differentiated cells. This patient presented 11 months later with ipsilateral axillary lymph node metastasis, which emphasizes the unfavorable prognosis of this synovial sarcoma variant. The indolent growth pattern of this sarcoma justifies the well circumscribed initial stages, which progressively infiltrate adjacent structures with lung metastasis (80%) and lymph node involvement (20%) and thus corroborates the importance of early diagnosis and proper treatment.
Keywords: Pathology; Sarcoma, synovial; Soft tissue neoplasms

INTRODUCTION

Synovial sarcomas (SS) are rare benign neoplasms that encompass from 5 to 10% of soft tissue sarcomas.[1,2] They affect mainly young adults and present as a painful slow growth mass located in deep soft tissues of extremities, near joints, even though their onset can be in any part of the body. Primary involvement of the epidermis in these sarcomas is extremely rare.[1] This report illustrates a poorly differentiated synovial sarcoma in the wrist with extension to the skin and with unfavorable evolution characterized by ganglionary and pulmonary metastases.

CASE REPORT

Male patient, 34 years old, with history of painful lesion for 3 months, ulcerovegetative, sessile, infiltrating the dermis for 3 cm on its greater axis, in the right wrist (Figure 1). A physical examination did not show either lymphadenomegalies or movement limitation of the limb. Excision of lesion was performed and histopathological study requested. In the histological sections stained by hematoxilin-eosin a malign neoplasm it was evidenced, with proliferation of poorly differentiated cells with scarce, irregular and eosinophilic cytoplasm, hyperchromatic round nuclei and conspicuous nucleoli suggesting poorly differentiated SS (Figures 2 and 3). The immuno-histochemical technique revealed focal positivity for epithelial membrane antigen (EMA) and TLEI (Wnt pathway, synovial sarcoma) and negativity for cytokeratins (AE1/AE3), emphasizing a poorly differentiated profile of this SS (Figure 4). The patient returned after 11 months with a painful tumor measuring 9x9 cm, erythematous, of stony consistency in the right axillary region suggesting lymph node metastasis (Figure 5). He presented considerable weight loss, intermittent fever and limitation of wrist movement. Physical examination did not show other lymphadenomegalies. During diagnosis the thorax CT scan detected a poorly defined nodule, spiculated margins, dense soft parts and calcification foci in the right lower lobe with 1 cm and another similar subpleural nodule in the middle lobe with 0.4 cm suggesting pulmonary metastasis. Patient in stage IV (any G, any T, N1M0 or M1) was referred to the clinical oncology department for treatment of pulmonary and lymph node metastasis.[3]

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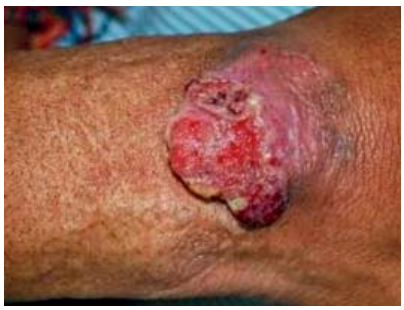

Figure 1: Wide base ulcerovegetative lesion, sessile, painful, infiltrating the dermis measuring 3 x 2.7 cm in right wrist. Front view

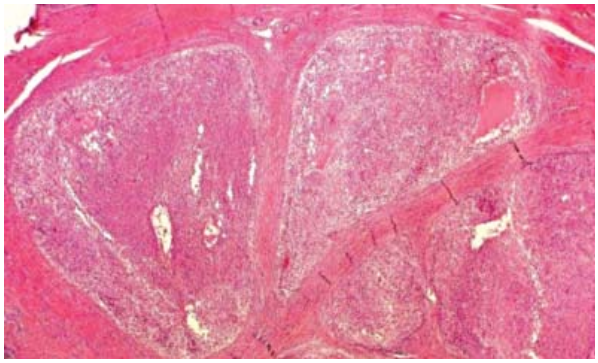

Figure 2: Fragment of cutaneous biopsy stained with hematoxilin-eosin 100x evidencing multiple nodular formations in the dermis formed by proliferation of neoplastic cells separated by streams of connective tissue

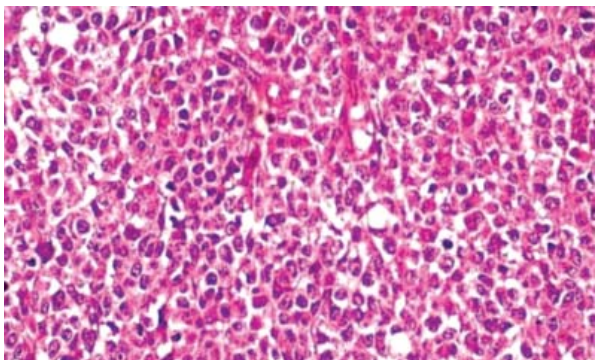

Figure 3: Fragment of cutaneous biopsy stained with hematoxilin-eosin 100x evidencing poorly differentiated cells with scarce, irregular and eosinophilic cytoplasm with hyperchromatic round nuclei and conspicuous nucleoli

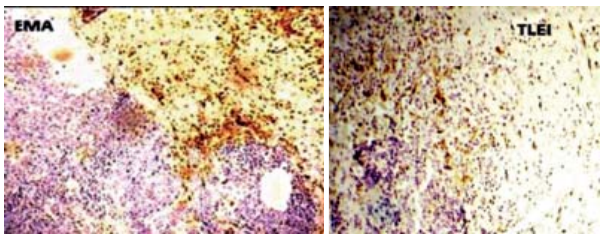

Figure 4: Immunohistochemistry of cutaneous biopsy focally positive for epithelial membrane antigen (EMA) and TLEI (Wnt pathway, synovial sarcoma)

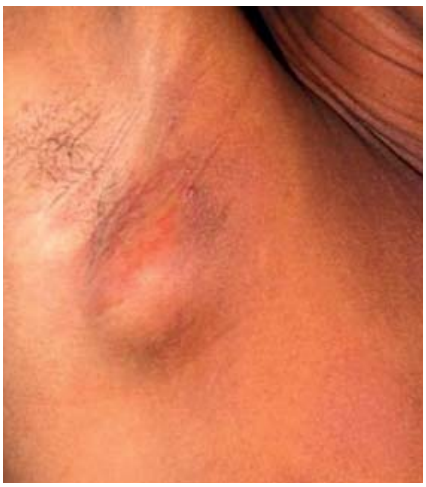

Figure 5: Painful tumor measuring 9x9 cm, erythematous, of stony consistency, not adhered to deep planes in the right axillary region suggesting lymph node metastasis

DISCUSSION

SS are malignant rare tumors that affect mainly young adults. For this reason, early treatment has an important impact on life expectancy.[1,2,4] They are located in deep soft tissues, close to the joints, especially in the lower limbs, being rarely located in the wrist.[1,2,3] In initial stages the tumor is well defined, but gradually infiltrates adjacent structures and/or involves more remote sites like the lungs (80%) and lymph nodes (20%) by metastasis.[2,3] The diagnosis depends on histopathological study that can identify 4 subtypes: biphasic, monophasic epithelial, monophasic fibrous and the poorly defined SS. Many times the histology of these tumors looks like metastatic carcinoma, malignant melanoma, epithelioid sarcoma and malignant schwannoma raising the need of immunohistochemistry for differential diagnosis.[1,2] Almost all SS are positive for vimentin, cytokeratin EMA and TLEI (Wnt pathway, synovial sarcoma). In the poorly differentiated form the focal marker of EMA and TLEI on neoplastic cells is more frequent than that of cytokeratins, which is usually negative, confirming the immuno-histochemical panel finding of the report.[1,5] The cytogenetic study helps in the diagnosis of non-characteristic cases. Over 90% of the SS present translocation between chromosome 18 and X, t(X; 18) (p11; q11) arising from the fusion of gene SYT with the gene SSX1 (biphasic) or SSX2 (monophasic).[1,5] The biological behavior of SS is variable, with several factors negatively changing the prognosis, as poorly differentiated histological subtype, tumoral dimension > 5 cm, lymph node involvement and presence of metastases (mainly pulmonary).[3,6] These factors compose TNM staging criteria and direct the treatment for each stage of the disease. The low incidence of SS and the diverse forms of treatment in different centers makes the evaluation of effectiveness of therapeutic means used more difficult.[2,3] Like in other sarcomas, the stan-

dard treatment is wide surgery margins, as performed in this case. Due to lack of random studies that indicate an adequate margin for resection an arbitrary value of 2 cm is used.[3,5,6] In spite of the use of wide resections, local or remote relapses of these tumors are common. In the case studied there was pulmonary and lymph node metastasis 11 months after the onset of lesion and 8 months after tumoral resection, but there are reports of onset after 5 years. There is some consensus in indicating adjuvant radiotherapy for high degree tumors, proximal G2 or head and neck, although studies suggest that the practice diminishes local relapse, but does not affect survival in high degree tumors.[3,7] The role of chemotherapy is still uncertain, but currently SS have been considered chemosensitive and indications of its use have been increasing.[3,5] Post-treatment periodic monitoring is recommended in order to diagnose complications early and increase the life expectancy of these patients.[3,7] In this case we highlight the extension to the skin (extremely rare) and the unfavorable evolution with evidence of lymph node and pulmonary metastasis, diminishing the life expectancy of the patient. ❑

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