ISSN-e 1984-8773



Surgical & Cosmetic Dermatology

www.surgicalcosmetic.org.br/

Fibromyxosarcoma mimicking a benign cystic lesion: a case report of a high-grade mesenchymal tumor in an atypical site

Fibromixossarcoma mimetizando lesão benigna cística: relato de caso de tumor mesenquimal de alto grau em localização atípica

DOI: http://www.dx.doi.org/10.5935/scd1984-8773.2024160305

ABSTRACT

Fibromyxosarcoma is a rare subtype of soft tissue sarcoma characterized by the presence of spindle cells and myxoid matrix. We report the case of an older patient who presented with progressive growth of a nodule in the interscapular region, and further investigation confirmed the diagnosis of high-grade fibromyxosarcoma. Notably, this neoplasm more commonly occurs in the lower limbs and may exhibit recurrence associated with increased histological grade. Therefore, early diagnosis and treatment are crucial for mitigating morbidity and mortality. We emphasize the significance of diagnosing an atypical case and providing timely therapeutic intervention, contributing to the optimization of clinical outcomes. Keywords: Sarcoma; Neoplasms, Connective and Soft Tissue; Solitary Fibrous Tumors

RESUMO

O fibromixossarcoma é um subtipo raro de sarcoma de partes moles, caracterizado pela presença de células fusiformes e matriz mixoide. Relatamos o caso de uma paciente idosa que apresentou crescimento progressivo de um nódulo interescapular, cuja investigação confirmou o diagnóstico de fibromixossarcoma de alto grau. É notório que esta neoplasia ocorre mais comumente nos membros inferiores e pode exibir recorrência associada ao aumento do grau histológico. Por isso, diagnóstico e tratamento precoces são fundamentais na mitigação da morbimortalidade. Enfatizamos a relevância do diagnóstico de um caso atípico e da intervenção terapêutica oportuna, contribuindo para a otimização dos desfechos clínicos.

Palavras-chave: Sarcoma; Neoplasias de Tecido Conjuntivo e de Tecidos Moles; Tumores Fibrosos Solitários

Case Report

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Financial support: None. Conflicts of interest: None.

Submitted on: 20/09/2023 **Approved on:** 22/10/2023

How to cite this article:

Souza CA, Motta IP, Azevedo RL, Amstalden EMI, Buffo TH. Fibromyxosarcoma mimicking a benign cystic lesion: a case report of a high-grade mesenchymal tumor in an atypical site. Surg Cosmet Dermatol. 2023;15:e20230305.



INTRODUCTION

Fibromyxosarcoma is a rare subtype of soft tissue sarcoma, composed of spindle cells and myxoid stroma. It is the most common mesenchymal neoplasm in older patients, presenting as painless, slow-progressing nodules, common in the lower extremities.1 Histologically, according to cellularity and atypia, they can be classified as low, intermediate, or high grade. This classification has no correlation with the high risk of recurrence (between 50-60% for all subtypes), but it is a predictor of metastases, which are present in 20-35% of intermediate and high grades, and are uncommon in low-grade sarcomas.² Unlike other sarcomas, myxofibrosarcomas tend to gradually present with a higher histological grade with each recurrence.3 This highlights the importance of early diagnosis, staging, and treatment, with a view to lower morbidity and mortality. We report the case of an older patient with high-grade myxofibrosarcoma in the interscapular region.

CASE REPORT

A 68-year-old female patient presented with a one-yearold, painless nodular lesion in the interscapular region. The initial dermatological physical examination revealed a painless, mobile nodular lesion measuring 6×4 cm. She had a previous soft tissue USG at the beginning of the condition, compatible with an epidermal cyst with free muscle planes (Figures 1A-1B). As the clinical picture was not consistent with the imaging exam, a CT scan was requested, which showed that it was a 7×3 cm cystic lesion, and excision was scheduled. Following incision, amorphous, gelatinous, blackened tissue could be seen, not corresponding to the initial hypothesis (Figures 1C-1D). A deep biopsy of the lesion was then performed, with histo-



FIGURE 1: A-B - Nodular lesion in the interscapular area prior to biopsy. C: Appearance of the lesion after the biopsy, showing dehiscence of the sutures. D: Exuberant exophytic growth of the tumor mass can be seen, and also the infiltrative aspect of the muscle planes

pathological results showing a high-grade myxofibrosarcoma (Figure 2). Magnetic resonance imaging of the spine was requested, showing the presence of an expansive lesion measuring 10.1 \times 8.8 \times 4.0cm, invading the muscle bulge of the underlying trapezius, with no signs of invasion of other muscle planes or adjacent bone structures (Figure 3). For staging, a PET-CT scan was performed, which showed hypermetabolism in bilateral axillary lymph nodes, suggestive of metastases, not confirmed through biopsy of the hypercapturing lymph nodes (Figure 4). The patient was referred to the Orthopedics team and the lesion was excised with wide margins. The patient was then referred to Oncology for clinical follow-up, and no adjuvant treatments were indicated. The patient is being followed up by a multi-disciplinary team six months after excision, with no recurrence (Figure 5).

DISCUSSION

Fibromyxosarcoma is a rare subtype of soft tissue sarcoma, first described by Enzinger and Weiss in 1977.¹ Previously called malignant fibrous histiocytoma myxoid variant, it has a mesenchymal origin and is composed of spindle cells and myxoid stroma. It is the most common malignant mesenchymal neoplasm in older patients, predominantly affecting males in the 50s to 70s.¹⁻³

The most common site is the lower limbs, with lesions rarely occurring in the trunk, upper limbs, or cephalic segment.



FIGURE 2: Neoplasm showing tissue with atypical and pleomorphic fibroblastic proliferation, sometimes containing binucleation or multinucleation, hyperchromatism, cytoplasm varying from slightly acidophilic to more abundant and foamy, also called pseudolipoblasts (red arrow). Note the presence of atypical mitotic figures (blue arrow). The stroma consists of abundant and richly vascularized myxoid tissue, with delicate, curved vessels. Note the relationship between the tumor and the muscular fascia (*).



FIGURE 3: MRI: Expansive lesion, measuring 10.1 × 8.8 × 4.0cm, with invasion of the underlying trapezius muscle bulge



FIGURE 5: Scar appearance six months after surgical excision



FIGURE 4: PET-CT scan: Heterogeneous interscapular mass, compatible with primary neoplasm. Hyper-uptake of axillary lymph node level I on the left and level V bilaterally

In general, it presents as painless, slow-progressing, normochromic, or slightly erythematous nodules or tumors. It occurs more frequently in subcutaneous soft tissues than in deep tissues.^{1,4,5}

Histopathological examination is the gold standard for diagnosing the disease. The sample must be deep to guarantee a reliable assessment of the material. Superficial biopsies can show benign characteristics or even subclassify a high-grade tumor.⁶ Histologically, it is classified as low, intermediate, or high grade, according to cellularity and atypia. Immunohistochemistry shows low specificity in myxofibrosarcoma and is generally positive for vimentin and rarely for smooth muscle actin, S100, and desmin.^{6,7}

The treatment of choice is excision of the lesion with wide margins. Local recurrence occurs in 50–60% of cases, and this risk is apparently not related to the depth of the lesions or the histological grade.⁸ However, there is a tendency for the myxofibrosarcoma to gradually become more cellular, more pleomorphic, more mitotically active, and thus have a higher histological grade with each recurrence. This characteristic is not observed in other sarcomas. Metastases are rare in low histological grade tumors, but occur in 20–35% of intermediate and high grade tumors, especially to the lungs and bones. The differential diagnosis includes other myxoid tumors.^{1,2,8}

The ability of the tumor to present high rates of recurrence and advance in histological grade supports the need for early diagnosis and treatment, so as to reduce morbidity and mortality.

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Statistical analysis; approval of the final version of the manuscript; study design and planning; preparation and writing of the manuscript; data collection, analysis, and interpretation; active participation in research orientation; intellectual participation in the propaedeutic and/or therapeutic conduct of the cases studied; critical literature review; critical review of the manuscript.

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