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Case Report

Benign ossifying fibromyxoid tumor: A case in disguise

Soumya Kori[1]*, Shwetha Ramu[1], HR Chandrashekar[1]

Dept. of Pathology, J.J.M Medical College, Davangere, Karnataka, India

Abstract

Introduction: This case report documents a rare instance of ossifying fibromyxoid tumor (OFMT), a soft tissue tumor with uncertain differentiation, in a 79-year-old female. The patient presented with a progressive, painless swelling in the posterior right thigh. Ossifying fibromyxoid tumor (OFMT) is an uncommon soft tissue neoplasm characterized by variable malignant potential (Enzinger et al., 1989). With fewer than 300 reported cases, OFMT primarily affects adults, with a slight male predilection.

Case Report: A 79-year-old female presented with a 12-month history of progressive, painless swelling in the posterior right thigh, measuring 12 x 10 cm. Diagnostic Evaluation:- Fine needle aspiration cytology (FNAC): Suggested epidermal inclusion cyst. Radiographic examination: Soft tissue swelling with focal calcification and intact cortical bone. Surgical excision was performed, and histopathological examination revealed a capsulated tumor comprising epithelioid cells within a fibromyxoid matrix, surrounded by woven and lamellar bone. Immunohistochemical Analysis: - Desmin: Moderately positive and S-100: Positive. The histopathological and immunohistochemical findings confirmed the diagnosis of OFMT. This case highlights the importance of considering OFMT in the differential diagnosis of soft tissue tumors.

This case report is presented for its rarity.

Keywords: Soft Tissue Tumour, Ossifying fibromyxoid tumor, S-100

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1. Introduction

OFMT was initially characterized in 1989 by Enzinger et al. through an analysis of 59 distinct histological cases.[1] The World Health Organization (WHO) classifies OFMT as a tumor of uncertain differentiation, exhibiting variable malignant potential.[2] Demographics and Clinical Features: OFMT affects adults across various age groups (14-83 years), with a median age of approximately 50 years. Males are slightly more affected than females (1.5:1 male-to-female ratio). Tumors commonly arise in proximal limbs, limb girdles, distal upper and lower limbs, head and neck, and trunk. Subcutaneous tissues are primarily involved, but deep muscle and bone sites can also be affected. Typical presentations include slow-growing, painless masses or palpable nodularity without associated pain. The clinical course ranges from 1 to 20 years (median: 4 years). Tumor sizes vary from 1 to 14 cm (average: 4-5 cm). Treatment and Prognosis: Most OFMTs are benign and treatable with excision. However, local recurrence occurs in 17% of cases. Malignant OFMTs comprise 5% of cases, with a 60% metastatic potential.[1] The cellular origin of OFMT remains unknown. However, neuroectodermal origin is suspected based on cytoarchitectural ndings and positive staining for markers like S100, desmin, CD56, CD99, and neurofilament

2. Case Report

2.1.1. Clinical history and examination

A 79-year-old female farmer presented with a painless, progressively enlarging swelling in the posterior aspect of her right thigh, spanning 12 months. The swelling measured 12 x 10 cm. Systemic Examination: No significant findings were observed. Radiological Examination: X-ray of the right femur revealed: - Soft tissue swelling with focal calcification on the posterior aspect of the proximal femur, Intact bony

*Corresponding author: Soumya Kori
Email: soumyakori4@gmail.com

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cortex - No bony erosion. Further Diagnostic Evaluation: Fine needle aspiration cytology (FNAC) was performed, and the aspirate was sent for Cytology. Culture and sensitivity testing results were negative. This presentation suggests a soft tissue tumor, potentially an OFMT, given the - Painless swelling - Gradual progression and Focal calcification. Further histopathological examination and immunohistochemical analysis would confirm the diagnosis.(**Figure 1**)

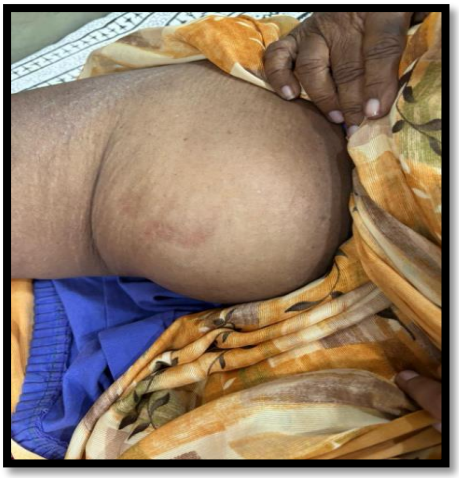

Figure 1: Clinical image demonstrating a 12 x 10 cm soft tissue mass in posterior aspect of right thigh

2.1.2. Gross examination

Received skin attached soft tissue mass measuring 9X8X5 cm. External surface -skin attached on one side. Cut surface showed a lobulated grey white tumour, firm in consistency with foci of calcification and areas of mucoid change.(**Figure 2**)

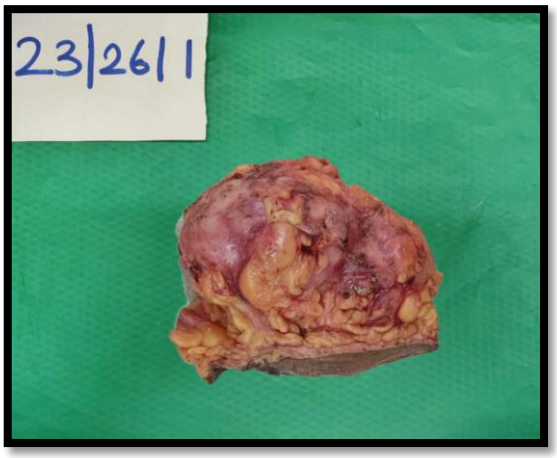

Figure 2: Soft tissue mass measuring 9x8x5 cm. External surface – skin attached on one side. On cut surface- lobulated grey white tumor

2.1.3. Microscpic examination

Microscopic examination showed capsulated tumor with tumor cells arranged in lobules, sheets and nests along with fibrohyaline and myxoid stroma. These cells are round to spindle having clear to pale cytoplasm, bland nuclei and fine chromatin. Occasional mitosis seen (<2/50Hpf). Also seen is significant peripheral shell of woven and lamellar bone (ossification). There are seen branching capillaries in the stroma. However, no increased cellularity, cytologic atypia or necrosis is seen.(**Figure 3**, **Figure 4**, **Figure 5**, **Figure 6**, **Figure 7**)

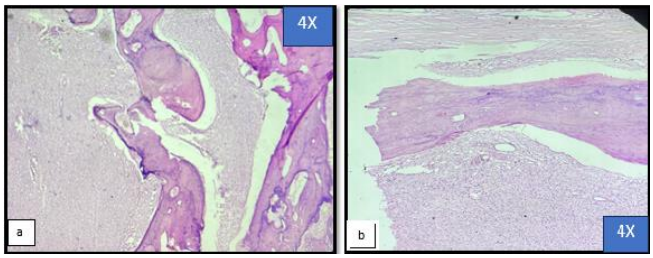

Figure 3: a,b: Capsulated tumor with peripheral shell of woven and lamellar bone

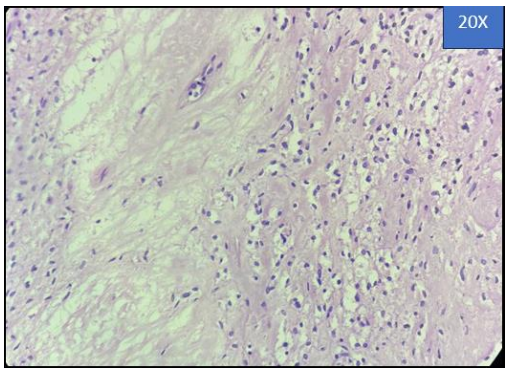

Figure 4: The cells are dispersed in cords , nests, sheets, lace-like patterns or singly in patternless distributions, and are surrounded by a variably fibromyxoid matrix .

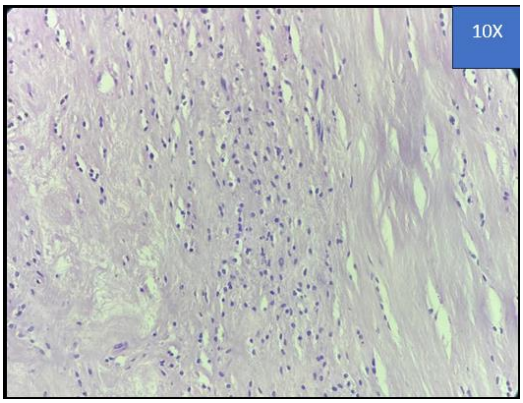

Figure 5: Proliferation of tumor cells embedded in fibrohyaline stroma.

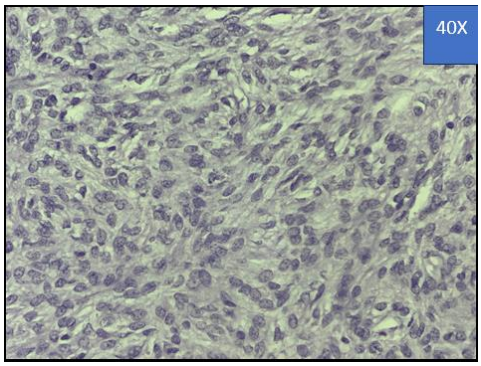

Figure 6: Bland oval to round nuclei with fine chromatin and smooth nuclear borders. The cytoplasm is palely eosinophilic, ill defined, and delicate with amphophilic to clear. Occasional mitosis seen (<2/50Hpf)

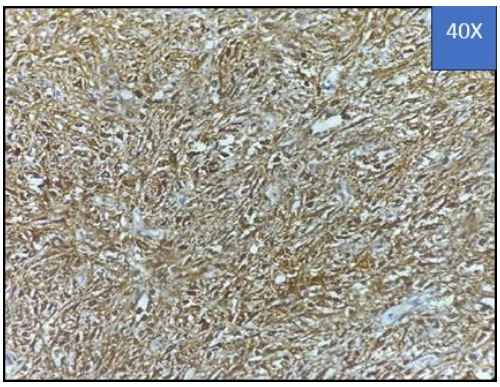

Figure 7: Strong S100 expression in neoplastic cells.

3. Discussion

Enzinger et al. first described OFMT as a distinct soft tissue neoplasm characterized by well-circumscribed nodular masses in subcutaneous tissues or muscles.[1] Classification and Risk Stratification: Folpe and Weiss (2003) proposed a risk stratification system categorizing OFMTs as:- Typical: Low nuclear grade, cellularity, and mitotic activity (2 MF/50 HPF), while malignant ones have mitotic activity of >2 MF/50 HPF, high cellularity ,nuclear grade ,and these cases found to be associated with distant metastasis. The atypical types do not meet the criteria of either of above.[2] Immunohistochemistry: OFMTs exhibit S100 protein expression, more prominent in typical subtypes. Other positive stains include: - Desmin Smooth muscle actin, Pancytokeratin Epithelial membrane antigen. Differential Diagnosis: - Solitary fibrous tumor, Sclerosing epithelioid fibrosarcoma, Malignant peripheral nerve sheath tumor, Synovial sarcoma Ossifying hematoma, Ossifying epithelioid hemangioendothelioma. Molecular Studies: - Novel EP400-PHF1 fusion gene on chromosome 6p21 is associated. PHF1 gene rearrangement in 80% of OFMTs and EP400 fusion in 44% of cases noted.[5] Treatment and Prognosis: - Surgical excision is the primary treatment. Follow-up recommended, especially for atypical and malignant types, due to local recurrences and distant metastases. 5-year survival rates: 59-100% and 10 year survival rates: 40-89%.[6,7,8]

4. Conclusion

Ossifying bromyxoid tumor (OFMT) is a rare neoplasm with variable biological behavior characterized by intermediate biological potential. Typically arising in the extremities or trunk, OFMT exhibits: - Uniform polygonal cells within fibromyxoid stroma and peripheral rim of lamellar bone seen. However, OFMT's histologic spectrum and morphologic overlap with other soft tissue neoplasms can complicate diagnosis. Accurate diagnosis is crucial due to: Low-grade morphology with potential for recurrence and metastasis. Diagnostic Con rmation using combination of morphology and immunohistochemistry i.e coexpression of S100 protein and desmin (observed in a minority of cases). Establishing the correct diagnosis ensures appropriate management, as most OFMTs behave benignly, while others carry a risk of local recurrence and metastasis

5. Source of Funding

None.

6. Conflict of Interest

None.

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