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CASE REPORT

Primary Osseous Fibroblastic Reticular Cell Tumour of the Proximal Humerus: A Rare Case Report and Review of Pathogenesis

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Abstract

Abstract

Fibroblastic reticular cell tumour (FRCT) is an exceptionally rare neoplasm arising from specialised fibroblastic reticular cells of the lymphoid stromal network. Primary involvement of bone is extraordinarily uncommon, with only isolated cases reported in the literature. We present a case of a 40-year-old woman with a destructive lesion of the proximal humerus subsequently diagnosed as FRCT. The patient was treated with neoadjuvant chemotherapy followed by wide surgical excision and endoprosthetic reconstruction. This report highlights the clinical, radiological, pathological, and therapeutic challenges associated with this rare entity and reviews emerging concepts of FRCT pathogenesis, including ectopic stromal differentiation, chemokine-driven microenvironmental mimicry, and mismatch-repair deficiency. This case contributes to the limited body of literature on primary osseous FRCT and underscores the importance of multidisciplinary management.

Keywords: Oncology, Fibroblastic, Bone Tumour, Humerus

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INTRODUCTION

Fibroblastic reticular cell tumour (FRCT) represents one of the rarest neoplasms within the spectrum of haematolymphoid stromal tumours. These tumours originate from fibroblastic reticular cells (FRCs), specialised mesenchymal stromal cells that reside predominantly in lymph nodes, thymus, and spleen. FRCs play a critical role in maintaining lymphoid architecture and regulating immune cell trafficking through the secretion of chemokines such as CCL19 and CCL21 (1).

Because FRCs are not native constituents of osseous tissue, primary FRCT arising within bone is exceptionally rare. Most reported cases involve lymph nodes or spleen (2), with only sporadic reports of extranodal disease affecting the oral cavity, breast, and spine (2-4). Owing to its rarity and histomorphological overlap with spindle cell sarcomas and other dendritic cell neoplasms, accurate diagnosis requires meticulous correlation of histopathological, immunohistochemical, and molecular findings.

In the absence of standardised treatment guidelines, management strategies for FRCT are largely extrapolated from soft tissue sarcoma and dendritic cell tumour paradigms, with wide surgical excision forming the cornerstone of therapy. We report an exceptionally rare case of primary FRCT of the proximal humerus and discuss its proposed pathogenesis, diagnostic pitfalls, and therapeutic considerations.

CASE REPORT

A 40-year-old right-hand-dominant Malay woman presented with a three-month history of progressive right shoulder pain associated with increasing swelling and functional limitation. The pain was dull, persistent, and exacerbated by activity. She denied constitutional symptoms, trauma, or previous malignancy.

Physical examination revealed a firm, tender mass over the proximal humerus with restriction of both active and passive shoulder movements secondary to pain. Distal neurovascular examination was unremarkable.

Magnetic resonance imaging (MRI) demonstrated an expansile intramedullary lesion involving the proximal humerus, associated with cortical destruction and extra-osseous soft tissue extension. The lesion exhibited heterogeneous T1- and T2-weighted signal intensities with areas of necrosis and heterogeneous contrast enhancement.

Ultrasound-guided core needle biopsy revealed a spindle-cell neoplasm arranged in intersecting fascicles and whorled patterns, with moderate nuclear atypia and focal tumour necrosis. Immunohistochemistry showed diffuse positivity for vimentin and smooth muscle actin (SMA), with negative staining for S100, CD21, CD23, and CD35. Based on the morphological features, immune-profile, and exclusion of other spindle cell sarcomas and dendritic cell tumours, a diagnosis of fibroblastic reticular cell tumour was established following multidisciplinary tumour board review and reference to established diagnostic criteria (5).

Given the locally aggressive radiological features, including cortical breach and tumour necrosis, neoadjuvant chemotherapy based on soft tissue sarcoma protocols was initiated. After three cycles, interval imaging demonstrated a partial radiological response with reduction in tumour bulk. The patient subsequently underwent wide surgical excision and endoprosthetic reconstruction of the proximal humerus.

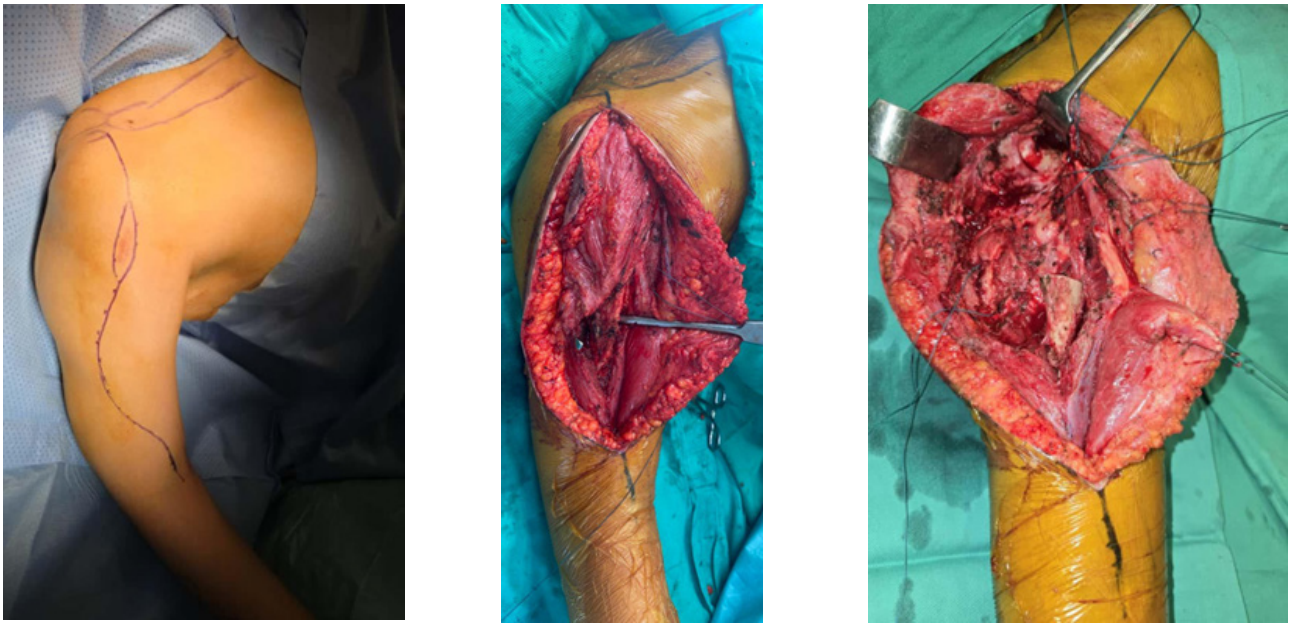

Figure 1: (A): Surgical Approach (B) & (C): Intraoperative findings

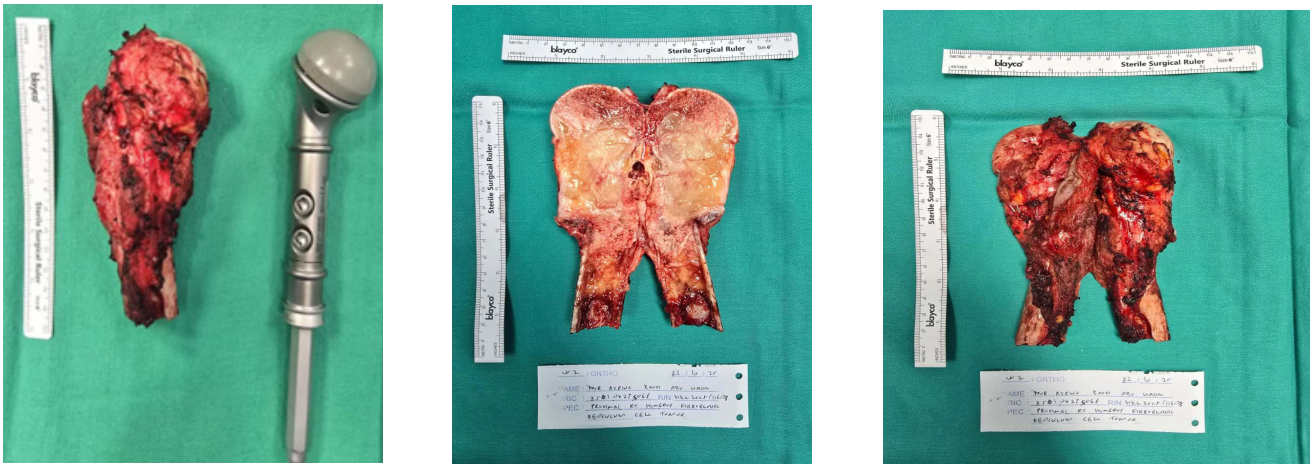

Figure 2: Intraoperative findings (A) Resected tumour with replacement prosthesis (B)(C) Tumour sample halved in both sides

Histopathological Findings

Histological examinations as shown in low and high power magnification revealed mostly hyalinized stroma with reactive and entrapped mature bone trabecular within the fibrotic matrix. Sheets of foamy macrophages and focal foreign body- type of giant cell reaction were identified. No residual viable malignant cells were seen.

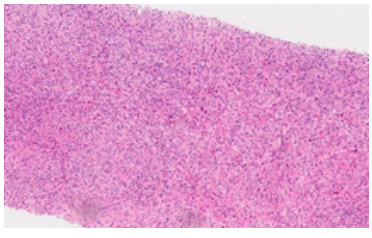

Figure 3. Hematoxylin and eosin staining (H&E) (Low power: magnification X4)

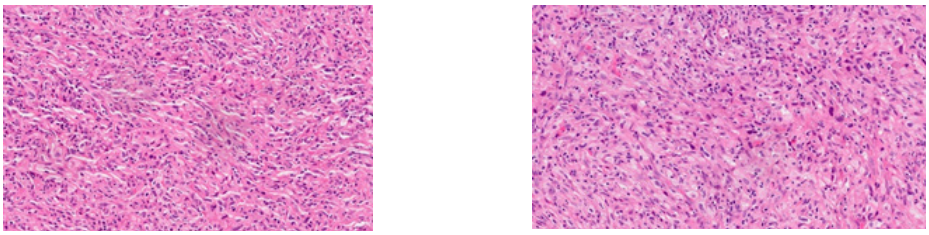

Figure 4. Hematoxylin and eosin staining (H&E) (High power: magnification X20)

DISCUSSION

Fibroblastic reticular cell tumour is an exceedingly rare neoplasm derived from fibroblastic reticular cells, which regulate lymphoid architecture and immune cell trafficking through chemokine expression, particularly CCL19 and CCL21 (1). The occurrence of FRCT in bone is biologically intriguing, as FRCs are not native to osseous tissue.

One proposed mechanism for primary osseous FRCT involves ectopic differentiation of bone marrow–derived mesenchymal stem cells into fibroblastic reticular–like cells under the influence of chronic inflammatory cytokines and aberrant stromal–immune signalling (6). Additionally, extranodal FRCTs appear capable of reconstructing lymphoid-like stromal microenvironments through autocrine chemokine expression, effectively mimicking their tissue of origin even in non-lymphoid sites such as bone (2,3).

Recent molecular studies have identified mismatch-repair (MMR) deficiency in a subset of FRCTs, suggesting a role for genomic instability in tumorigenesis and potentially explaining the variable and unpredictable biological behaviour of this tumour (7). Clinically, FRCT exhibits a wide spectrum of aggressiveness, ranging from indolent lesions to locally aggressive tumours with metastatic potential, most commonly involving the lungs and liver (2-4).

Given the absence of standardised treatment protocols, management is guided by extrapolation from soft tissue sarcoma principles. Wide surgical excision remains the mainstay of treatment, with chemotherapy or radiotherapy considered in selected high-risk cases, particularly in the presence of aggressive histological features, locally advanced disease, or metastasis (2). Risk stratification frameworks adapted from solitary fibrous tumour models, such as the Demicco system, incorporating tumour size, necrosis, depth, and mitotic activity, may assist in estimating malignant potential (8).

In the present case, the deep anatomical location, extensive cortical destruction, soft tissue extension, and tumour necrosis suggested an aggressive phenotype, supporting the use of neoadjuvant chemotherapy followed by wide resection and endoprosthetic reconstruction. This multimodal approach achieved satisfactory early oncological control while preserving limb function.

CONCLUSION

Primary osseous fibroblastic reticular cell tumour is exceptionally rare and poses significant diagnostic and therapeutic challenges. This case supports emerging concepts that fibroblastic reticular cells or their progenitors may undergo ectopic differentiation within bone, driven by inflammatory, chemokine-mediated, or genomic instability pathways. Complete surgical excision remains the cornerstone of treatment, with multimodal therapy reserved for selected high-risk cases. Continued reporting of cases with detailed molecular correlation is essential to advance understanding of FRCT pathogenesis, optimise management strategies, and refine prognostication.

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Data availability

The datasets generated during and/or analyzed during the current study are available from the corresponding author upon reasonable request.

Contributions

Research concept and design: NXY, EG, ATZY, OSL, SD

Data analysis and interpretation: EG, OSL, SD

Collection and/or assembly of data: EG, NXY, SD, ATZY,

Writing the article: EG, SD, ATZY, OSL, NXY

Critical revision of the article: NXY, EG, ATZY, SD, OSL

Final approval of the article: NXY, EG, SD, OSL, ATZY

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