Angiomyolipoma Distal Thigh with Osteonecrosis of Femoral Condyle – A Rare Tumour Entity

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INTRODUCTION:

Extra-renal angiomyolipoma (AML), a rare tumor classified as benign tumor of uncertain differentiation in the 2020 WHO Classification, usually manifests as a solitary renal tumor (80% of cases) and infrequently in the musculoskeletal system.[1] The cell of origin is unclear, with hypotheses proposing a cell resembling pericytes or primitive mesenchymal cells capable of differentiating into both smooth muscle and fat cells.[2]

REPORT:

A 40-year-old male, reported one-year history of left knee swelling and six months of localized medial knee pain, worsening with ambulation and knee movement, and accompanied by night and rest pain. Clinical examination revealed a localized soft tissue swelling of 6 cm x 6 cm with ill-defined borders and deep-seated characteristics. Radiographic showed a benign periosteal reaction with minimal cortical destruction, while MRI identified a soft tissue mass in the anteromedial surface of the left femur with intermediate intensity between muscle and fat, displacing the suprapatellar bursa and infiltrating the vastus medialis muscle with early osteonecrosis of underlying bone.

A Trucut biopsy revealed fibromyxoid changes and necessitating exclusion of low-grade fibromyxoid sarcoma. Wide resection of the left distal femur with endoprosthesis performed based on the provisional diagnosis of fibromyxoid sarcoma with bone infiltration.

The final histopathological examination (HPE) revealed angiomyolipoma with muscle infiltration and bony erosion, suggesting a locally aggressive benign lesion.

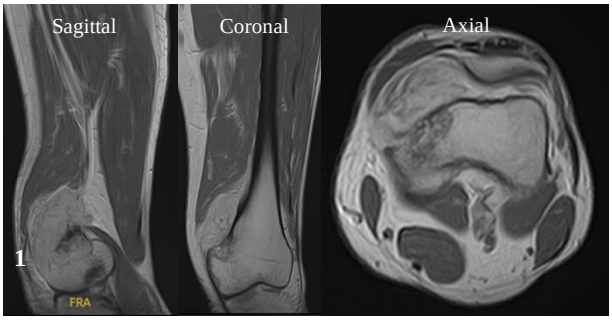

Figure 1: Contrasted MRI left thigh showed a contrast enhanced 2 cm x 4 cm x 8.1 cm soft tissue mass at the meta-diaphysis of the femoral anteromedial surface with bone early osteonecrosis (hypointensity).

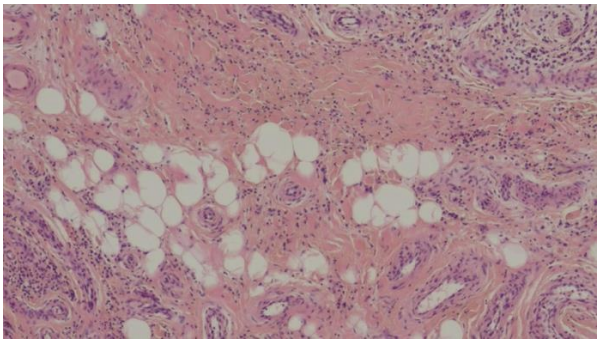

Figure 2: HPE showed mixture of blood vessels, adipocytes, and bundles of smooth muscle fibres in a fibrous background.

CONCLUSION:

This report highlights the diagnostic challenge of AML occurrences in the musculoskeletal system. Wide excision of angiomyolipoma is frequently curative.

REFERENCES:

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