Extraskeletal Osteosarcoma Mimicking Sclerosing Epithelioid Fibrosarcoma: A Case Report

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INTRODUCTION:

Extraskeletal osteosarcoma (EOS) is a rare variant of osteosarcoma with an incidence of less than 4% of osteosarcoma (1). Diagnosis is challenging due to its rarity, non-specific radiological features, and multiple histological mimickers (2). We report a rare case of quadriceps EOS which was initially identified as sclerosing epithelioid fibrosarcoma (SEF) based on the biopsy sample.

REPORT:

A 56-year-old lady presented with a painless enlarging mass over her right thigh. Examination revealed a deep-seated mass sized 15cm x 15cm located over the anteromedial aspect. Radiograph showed increased soft tissue shadow without calcification. MRI revealed a well-defined intramuscular soft tissue mass involving vastus intermedius and medialis with central necrosis. CT staging showed multiple lung nodules. A tru-cut biopsy identified SEF. She underwent wide resection involving vastus medialis, intermedius, and part of lateralis for wide margins. The exposed femur was covered with mobilized sartorius and adjacent quadriceps. The final histology indicated high-grade spindle cell sarcoma with osteoid formation, suggesting extraskeletal osteosarcoma. The lesion also includes hypocellular areas with predominantly sclerosing stroma, similar to the previous biopsy. The malignant cells are positive for SATB2 immunohistochemistry. All margins were negative for tumour cells. Following this, she underwent palliative chemotherapy.

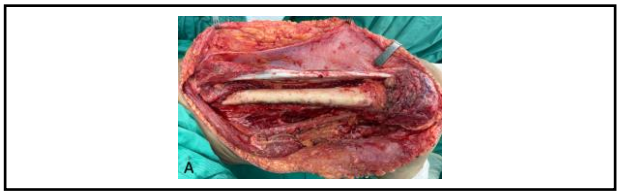

Fig.1: (A)After resection

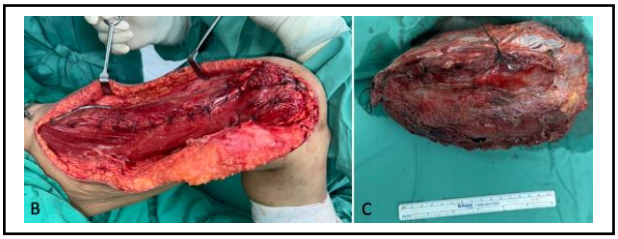

Fig.2: (B)Soft tissue coverage (C)Resected specimen

CONCLUSION:

An early and accurate diagnosis of EOS is crucial given its poor prognosis, with a survival rate of 58% (1). Some conditions that may be mistaken for EOS include sclerosing epithelioid fibrosarcoma, malignant peripheral nerve sheath tumour, and synovial sarcoma (2). Immunohistochemistry tests aid in diagnosis. Ultrasound-guided tru-cut biopsy or open biopsy are recommended for obtaining more representative samples. Treatment follows the management of conventional osteosarcoma. But, overall survival rates remain unchanged with this regime (2).

REFERENCES:

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