



LEIOMYOSARCOMA IN A PATHOLOGICAL FRACTURE OF PROXIMAL HUMERUS

Orthopaedic

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ABSTRACT

Leiomyosarcoma is an aggressive type of soft tissue neoplasm that originates from smooth muscle cells; it is typically found in the uterus, gastrointestinal tract, and retroperitoneum. Primary leiomyosarcoma of bone is an extremely rare phenomenon first reported in 1965. Here, we present a case of primary leiomyosarcoma of bone occurred in the proximal humerus of a 34-year-old man who was referred to our orthopaedic clinic with a chief complaint of right shoulder pain. Radiological investigations revealed a pathological fracture of right proximal humerus with a lytic lesion showing extensive periosteal reaction. Core needle biopsy of the lesion was performed, which pointed to primary leiomyosarcoma of bone as the underlying pathology.

KEYWORDS

INTRODUCTION-

Leiomyosarcoma is an aggressive type of soft tissue neoplasm that originates from smooth muscle cells. It is typically found in the uterus, gastrointestinal tract, and retroperitoneum. Primary leiomyosarcoma of bone is an extremely rare phenomenon, which was first reported by Evans and Sanerkin in 1965 (1). Due to its rare incidence and unspecific radiologic features, leiomyosarcoma of bone is commonly misdiagnosed with other pleomorphic undifferentiated sarcomas such as osteosarcoma, chondrosarcoma, or other unspecified sarcomas of bone (2). This pathology is predominantly reported in adult males, especially in the distal femur and other large bones. Here, we present a case of primary leiomyosarcoma of bone occurred in the proximal humerus of 34-year-old man.

Case Report-

A 34-year-old male presented to our out-patient department with acute onset of pain and inability to move the right shoulder following history of trivial trauma. On clinical examination, patient had tenderness over the proximal arm along with significant swelling of the right arm. There was a coarse crepitus palpable and abnormal mobility indicating the presence of a fracture and movements of the right shoulder was painful and restricted.

Radiographs showed a pathological fracture of the right proximal humerus with an eccentric lytic lesion, with a moth-eaten to permeative cortical destruction with periosteal erosions and wide zone of transition. An MRI was obtained to further define the extent and nature of the lesion, confirming the presence of a large ill-defined altered signal intensity heterogeneously enhancing lesion in the proximal one third of right humerus in the meta-diaphyseal region. The lesion showed mixed signal intensity on T2/STIR sequences with predominant hypointense areas and on T1WI the lesion showed hypointensity with endosteal scalloping. Periosteal soft tissue changes were also noted up to the capsule insertion in infero-medial aspect. Multiple enlarged ipsilateral axillary lymph nodes were visualized (Figure 1). Osteomyelitis, Bone cancer and metastatic tumours were considered the main differential diagnoses.

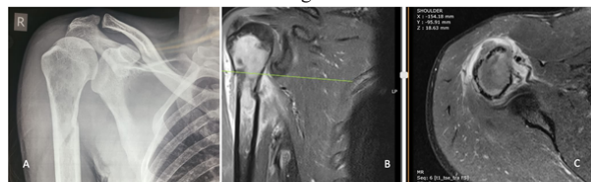


Figure 1- (A) X-ray showing Pathological fracture of the proximal humerus with an eccentric lytic lesion having moth eaten destruction and cortical erosions. (B & C) MRI showing heterogenous enhancing lesion in the meta-diaphyseal region of the proximal humerus with extensive soft tissue involvement.

The patient was admitted to the orthopaedic ward with preparation for core biopsy. Microscopic examinations showed bony trabeculae and tumour cells having elongated spindle nuclei with moderate amount of cytoplasm arranged in fascicular, storiform, and diffuse pattern with

areas of atypical mitosis favouring primary leiomyosarcoma of bone as underlying pathology (Figure 2). Subsequently, immunohistochemistry analysis revealed positive IHC markings for Vimentin, Desmin and Ki-67 (30-40%). IHC reports confirmed our diagnosis of primary leiomyosarcoma (Figure 3). Patient was planned for complete resection of the lesion and prosthetic replacement of the proximal humerus.

DISCUSSION-

Here we presented a rare case of primary leiomyosarcoma of the proximal humerus in a middle-aged man that was diagnosed incidentally with a pathological fracture. Leiomyosarcoma of bone is an uncommon bone neoplasm with a prevalence rate of less than 1% among all types of malignant bone tumours (3). By improvement of diagnostic tests and pathological examinations in recent years, the number of cases diagnosed with primary leiomyosarcoma has increased notably. Characteristic radiographic findings in primary leiomyosarcoma are osteolytic lesion with moth-eaten appearance and indistinct margins (2). Irregular cortical erosions may be apparent, but sclerosis and periosteal reaction are less common (3). Excisional biopsy and Immunohistochemical evaluations are often required for confirming the diagnosis (4). Major differential diagnoses of the leiomyosarcoma of bone are metastatic carcinoma of unknown primary origin, lymphoma, and osteosclerotic myeloma. It is important to differentiate between leiomyoma and leiomyosarcoma especially when a previous history of benign leiomyoma exists (5). Surgical resection with a wide margin is the main treatment option for primary leiomyosarcoma of bone. Therapeutic benefits of chemotherapy and radiotherapy are still unclear (3). Local recurrence of the tumour is uncommon, but the risk of distal metastasis is relatively high especially in patients with high-grade tumours (2). Tumour stage is the main predictor of long-term outcome, but it seems that the 5-year overall survival rate of primary leiomyosarcoma of bone is 59% that is similar to other common bone sarcomas (2).

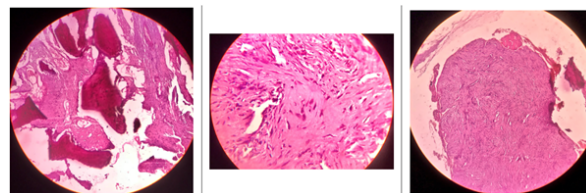


Figure 2- Microscopy showing bony trabeculae and tumour cells having elongated spindle nuclei with moderate amount of cytoplasm arranged in fascicular, storiform, and diffuse pattern with areas of atypical mitosis

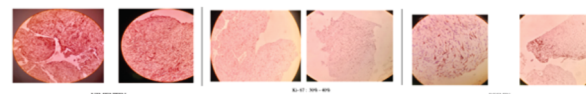


Figure 3- Immunohistochemistry analysis showing positive IHC markings for Vimentin, Ki-67 (30-40%) and Desmin.

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