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CASE REPORT

Management Challenges In Monophasic Synovial Sarcoma Of Distal Femur With Lung Metastasis: To Amputate Or Limb-Salvage Surgery

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INTRODUCTION

Synovial sarcoma is one of the commonest malignant soft tissue sarcomas with poor prognosis, especially when detected in late stages. It has a wide spectrum of biologic behavior, a high degree of local invasiveness and capability to spread trans compartment. There appears to be a lack in the reporting of these tumours locally as the challenges of diagnosis and management are often demanded at advance stages. We present a case of synovial sarcoma in an adolescent who presented very late with a huge lesion over right thigh with metastasis to the lung. This report highlights the diagnostic challenges which directly affect the type of surgical intervention and physical outcomes of patient.

CASE REPORT

A 16 year old female patient was referred to our clinic following the complaint of a mass over her right distal thigh for the past 2 years. The mass initially started as a small lesion, however, over a period of 4 months, its size increased progressively. Physical examination of the lesion demonstrates a 15 cm mass, which was tender, with normal overlying skin.

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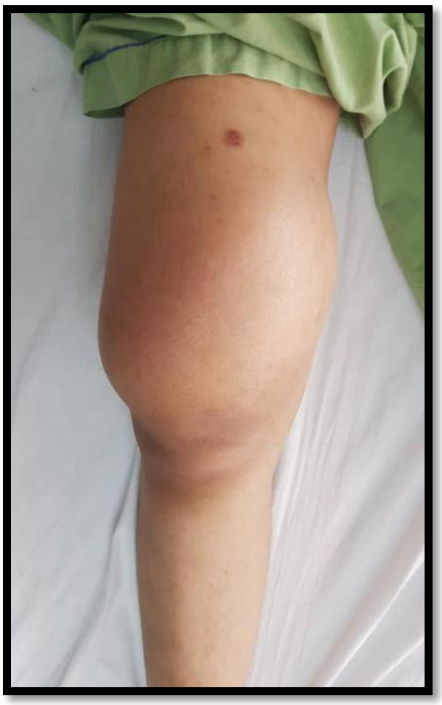


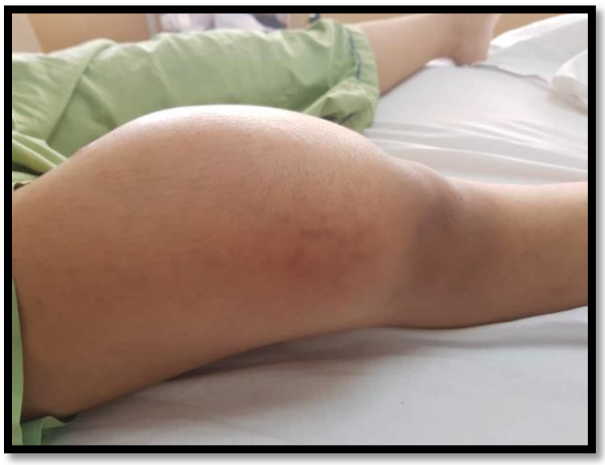
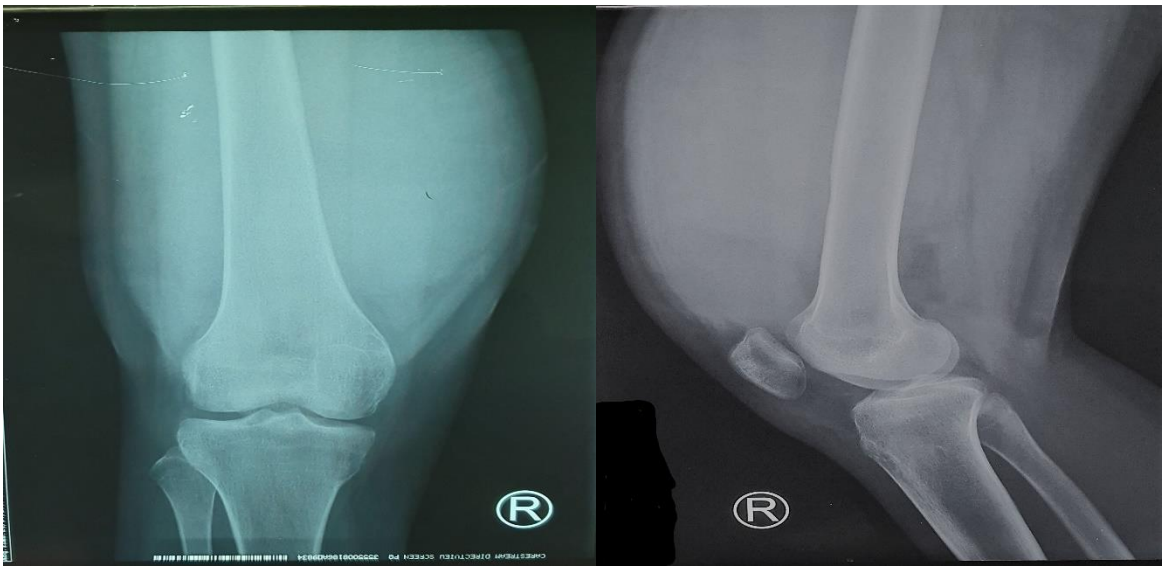
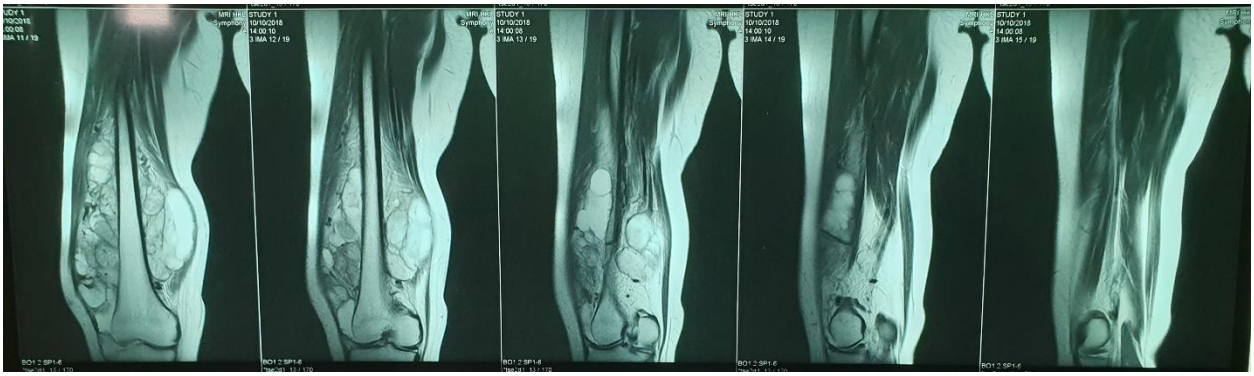


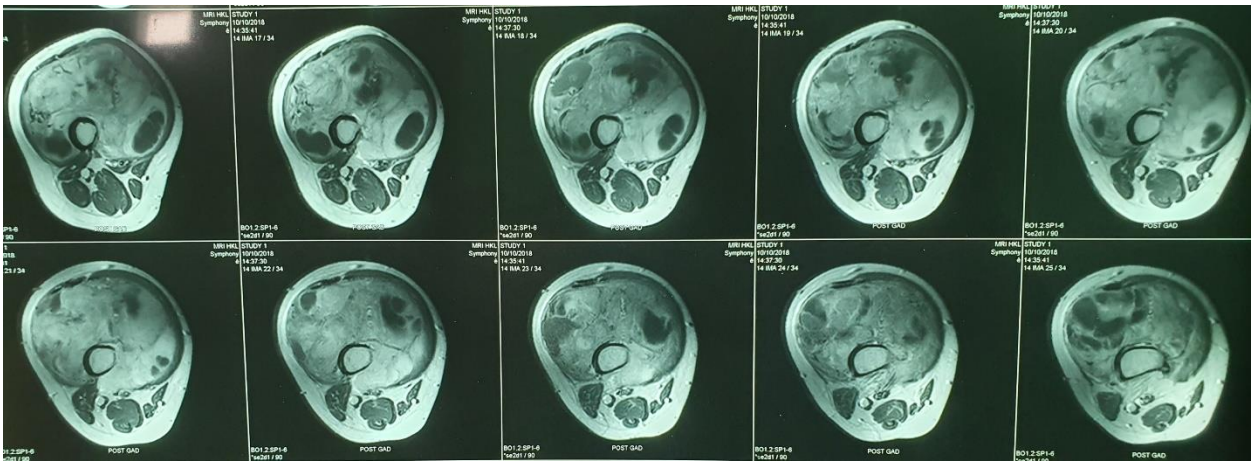
Fig 1 & 2:- Right Distal Thigh Mass

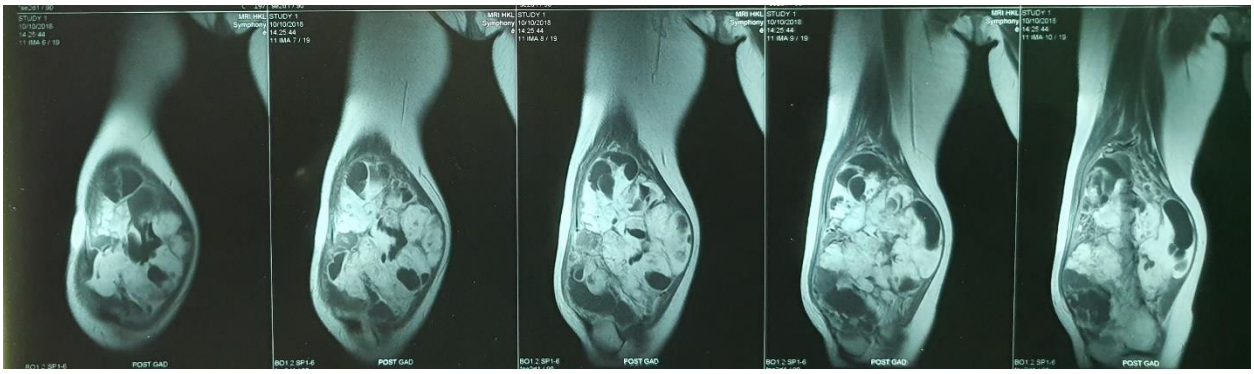
Right thigh revealed a soft tissue mass over the right distal thigh, measuring 8.9 x 11.5 x 17.0cm with normal underlying bones (Figure 1 & 2). MRI of the right distal thigh was done for the patient, which revealed a large, lobulated mass with its main bulk within the distal anterior compartment of the thigh. The mass is predominantly hypointense on T1W1 with heterogenous signal intensifies on T2W1. There is a heterogeneous enhancement post gadolinium contrast. The mass is encasing the distal femur, with no intramedullary extension.



Following the MRI, a trucut biopsy of the mass was done. Multiple strips of tissue from the mass were sent for histological evaluation. Histologically, the tumour consisted of spindle-shaped cells arranged in sheets, short fascicles, and pericytomatous pattern. These cells have ovoid, fusiform hyperchromatic nuclei with dispersed chromatin and inconspicuous nucleoli and indistinct cytoplasmic border. Immunohistochemistry evaluation shows the tumour cells were positive for TLE1, S100, CK7, EMA, and CKAE1/AE3, whilst negative for CD34 and Desmin. The diagnosis of monophasic synovial sarcoma of right distal thigh was made.







Contrast-enhanced computed tomography (CECT) was done for the patient which revealed 3 lung nodules scattered over bilateral lung fields and its ranging in size from 2 to 3.5mm, highly suggestive of lung metastasis. The patient was then subjected to 3 cycles of neoadjuvant chemotherapy using a regimen of Ifosfamide and Doxorubicin.

Repeat MRI post-neo-adjuvant chemotherapy, shows an increasing tumour size, measuring 9.3 x 12.6 x 19.0cm, with unchanged locoregional involvement of the lesion. The patient had undergone above-knee amputation. Intra-operatively, the cross-section of the tumour revealed multiple areas of necrosis and hemorrhage. Post-operatively, patient was subjected to adjuvant chemotherapy and radiotherapy, as well as rehabilitation following the above-knee amputation of right lower limb. Upon a 1-year followup, revealed a thankful patient ambulating with prosthesis and resumed studies in a local university.

DISCUSSION

Synovial sarcomas account for 7% to 10% of human soft-tissue sarcomas [1]. It arises at any age, but mainly affects young adults and more commonly males. Clinically, they appear as deep-seated slowly growing masses. In more than half of the cases, metastases develop, primarily to the lungs but also to the lymph nodes and bone marrow [1]. Synovial sarcomas occur most frequently in the lower and upper extremities, especially in para-articular regions and are associated with tendon sheaths, bursae, and joint capsules. Other frequent locations include the head, neck, and trunk, but, in rare cases, tumors have also been reported in the heart, lung, esophagus, small intestine, prostate, mediastinum, and retroperitoneum. The diversity of organs where synovial sarcomas arise contradicts its term. In fact, the term synovial sarcoma was coined in the first half of the 20th century to denominate tumours arising near joints which show a microscopic resemblance to synovial tissue. Later, consequent studies have demonstrated that synovial sarcomas display ultrastructural and immunohistochemical features of epithelial but not synovial differentiation [1]. Today it is accepted that synovial sarcomas are derived from unknown multipotent stem cells that are capable of differentiating into mesenchymal and/or epithelial structures.

On the basis of histopathological appearance, synovial sarcomas are usually divided into three subtypes: biphasic, monophasic, and poorly differentiated tumours [1]. Biphasic synovial sarcomas (BSS) are composed of two morphologically different cell types, epithelial and spindle cells, which vary in distribution, the latter being the larger component [3]. In contrast to BSS, monophasic synovial sarcoma (MSS) constitute only of spindle cells and is therefore morphologically similar to other spindle cell tumours such as fibrosarcomas.

Poorly differentiated synovial sarcomas (PDSS) encompass oval or spindle-shaped cells of small size, which look intermediate in appearance between epithelial and spindle cells [1]. Synovial sarcomas are often difficult to diagnose purely on histological and clinical grounds. In many cases, only with the aid of ultrastructural, immunohistochemical, or genetic studies is it possible to recognize these malignancies clearly. This is especially for MSS cases, the most frequent histological subtype, which could be misdiagnosed for spindle cell sarcomas, e.g., fibrosarcoma, leiomyosarcoma, malignant peripheral nerve sheath tumour (MPNST), hemangiopericytoma, or malignant fibrous histiocytoma [2].

Several studies have demonstrated that immunohistochemistry may be of help in the diagnosis of MSS since these tumours are generally positive for epithelial markers (e.g., cytokeratins and EMA) that are mostly absent in other soft-tissue sarcomas such as fibrosarcomas [3, 5]. Moreover, MSS cases are usually negative for muscle-associated markers (e.g., desmin and smooth-muscle actin), allowing distinction from leiomyosarcoma, and infrequently express S100 protein, which is present in the majority of MPNST cases [4, 5] PDSS may also be difficult to differentiate from other soft-tissue tumours, especially peripheral neuroectodermal and high-grade MPNST tumours. A recent

study indicates that PDSS may be immunohistochemically distinguishable from the former tumours based on the expression of the markers CD56 and CD99 and high-molecular-weight cytokeratins [3]. In contrast to other histological subtypes, BSS cases are clearly identifiable due to their characteristic biphasic morphology, which includes glandular or solid epithelial structures containing cuboidal or columnar cells, intermixed with a spindle cell component formed by densely packed elongated cells [4, 5].

Recent studies have shown that specific patient, tumour, and treatment factors influence the prognoses of synovial sarcoma patients. For example, young patient age, smaller tumour size, massive tumour calcification, numerous intratumoural mast cells, and distal location have all been reported to be favorable prognostic features [3]. In addition, poorly differentiated histology, tumour necrosis, vascular invasion, high mitotic rate, high proliferative index, DNA aneuploidy, and certain molecular genetic features have all been reported to be adverse prognostic factors [3]. Recent studies have claimed that adequate surgical margins and adjunctive chemotherapy and radiation therapy have improved prognosis compared with older series [2]. Unfortunately, many of the previous studies were too small to be subjected to multivariate statistical analysis.

Tumour size also strongly correlated with the outcome with the reason being; larger size suggests later tumour detection, may indicate rapid tumour proliferation, often correlates with a proximal location that would accommodate a larger tumour, and may cause more difficulty in achieving adequate surgical margins. Other studies of synovial sarcoma have also shown that tumour size is an independent prognostic factor for survival. The estimated 5-year survival for patients with tumours 5 cm was nearly 90%, compared with 40% for patients with tumours >5 cm. Distal tumour location has also been reported to be a favorable prognostic factor for patients with many different soft tissue sarcomas, including synovial sarcoma. The current standard treatment is wide resection followed by polychemotherapy with or without irradiation. Regional lymph nodes also should be removed. Neoadjuvant chemotherapy is a matter of debate. Initial surgical treatment with adequate surgical margins by surgeons experienced with sarcomas, preferably at specialized centers, should be considered to improve local control, outcome, and survival.

CONCLUSION

This report highlights the need for early detection and awareness of the referral of lumps and bumps to an orthopaedic oncologist. Late presentation very often leads to difficulties in management and physical outcomes. Younger patients with smaller tumours that are more distal in location have shown to have better prognosis. Wide resection with polychemotherapy with or without irradiation is the gold standard for treatment.

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