Unusual presentation of synovial sarcoma of Iliotibial band- A Case Report

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ABSTRACT

Synovial sarcoma is a rare soft tissue tumour which usually manifest as pain followed by swelling. We present two cases with unusual presentation of synovial sarcoma with limp with abduction deformity of hip.

Two cases of 7 year old boy and 13 year old girl presented identically with painful limp. Tenderness was present on anterolateral aspect above the knee joint and fixed abduction deformity of ipsilateral hip joint was noted. Bone scan was normal and MRI revealed soft tissue swelling arising from lateral aspect of distal thigh. On exploration, soft tissue tumor was arised from iliotibial band and biopsy revealed monophasic fibrous type of synovial sarcoma. Immunohistochemistry confirmed the same.

Orthopaedic surgeon must be aware about unusual presentations of symptoms and site of origin to prevent unnecessary delay in diagnosing rare highly malignant tumor.

Key words: Iliotibialband , Synovial sarcoma

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INTRODUCTION

Synovial sarcoma is most common non rhabdomyosarcomatous childhood soft tissue sarcoma. Usually synovial sarcoma presents with swelling with or without pain (1, 2, 3). Chotel et al highlighted the variability in presentation of this tumour and elusive nature of its diagnosis (4). Here we present two unique cases of synovial sarcoma of iliotibial band with unusual presentation.

CASE PRESENTATION

CASE -1: A 6.5 year old boy presented with limp for 1 year. Child walked with left side antalgic gait. Flexion and abduction deformity of left hip was noted. A painful hyperaesthetic area was marked in the lateral aspect of left knee. Radiograph of pelvis showed abduction

deformity with normal hip joints. Knee radiographs were normal. MRI scan showed pedunculated soft tissue arising superficial to lateral condyle and through iliotibial band. Bone scan showed normal uptake. Child underwent excision biopsy and was put on traction. Biopsy showed monophasic fibrous type of synovial sarcoma. Immunohistochemistry showed s-100+. Child later underwent 6 cycles of chemotherapy and radiotherapy. Pain and deformity were subsided following the treatment.

CASE -2: A 13 year old girl presented with painful limp since 1 year without history of trauma or constitutional symptoms. On examination, abduction deformity of left hip and lateral joint line tenderness in left knee was found. Radiographs of hip joint and knee joint were normal. MRI showed soft tissue tumour arising from under surface of iliotibial band. Bone scan was normal. Child underwent excision biopsy which showed as spindle cell sarcoma monophasic synovial sarcoma. Immunohistochemistry showed cd99+, vimentin +, bcl-2 +. The child was lost in follow-up.

DISCUSSION:

Synovial sarcoma is relatively rare form of soft tissue sarcoma (5,6). Synovial sarcoma accounts for approximately 1% of all childhood malignancy (7). Synovial sarcoma is the fourth most common soft-tissue sarcoma after malignant fibrous histiocytoma, liposarcoma, and rhabdomyosarcoma (1,8). Though called "synovial sarcoma", there is no convincing evidence that it always originates from synovial tissue, and less than 10% of these tumors occur intra-articularly (9). Usually arising in close proximity to tendon sheaths bursae and joint capsules, synovial sarcoma may also occur in the head and neck, abdominal wall and retro peritoneum (1).

The tumour can occur over a wide age range but usually is seen in adolescent and young adults of about 15 to 40 yrs age. Knee is the commonest large joint involved. The most common form of this disease is the biphasic type, with epithelial cells and fibroblast-like spindle cells. Synovial sarcomas, however, are unusual because they often do not present in a manner typical of soft-tissue sarcoma and delays in diagnosis are frequent (10). Most patients with soft tissue sarcoma present with relatively large mass and pain subsequently. Synovial sarcoma is considered to be a high grade tumor with a relatively poor prognosis.

The prognosis for 5-year survival ranges from 36% to 76%, and the 10 year survival rate is less (20% to 63%), because of late metastases."(1) Synovial sarcoma of Iliotibial band is a rare entity and with atypical presentation as in two of our cases makes the diagnosis difficult and thus delay in treatment. Early diagnosis and complete excision are associated with good survival for synovial sarcoma.

CONCLUSION

Soft tissue tumors usually pose challenge in their diagnosis with their varied clinical presentation. Treating surgeon must be aware of these presentations and always keep in mind about this malignant tumor whose early detection and treatment can add years to life.

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