Abstract

A 17 years old female patient presented to PMR OPD with fixed flexion deformity of left hip and knee and cachexia. Five years back a severe pain was suddenly developed in her left knee and thigh which was investigated for juvenile inflammatory arthropathy and rheumatic arthritis. At that time all the serological markers (ANA, RF, ASO titre) and x-ray of knee were normal. Subsequently left hip pain and restricted ROM were developed which made it clear that the knee pain was actually referred from hip. A plain x-ray of hip was done to rule out Perthe’s disease which was reported as avascular necrosis of femur.

When the patient was examined at PMR OPD, a CT scan of hip, routine hemogram, CXR, Mantoux test was advised considering a provisional diagnosis of infective pathology like TB hip with a differential of neoplasia in or around hip keeping in mind about cachexia and weight loss. Surprisingly CT scan showed a big mass originating from glutei muscles evading back of the thigh and even left sphincter ani muscle. Fortunately patient was continent at that time. Interestingly the pathological report suggested a relatively rare diagnosis which practically made the patient bedridden with commonly featured fixed flexion deformity.

Key words: Fixed flexion deformity, rehabilitation.

Introduction:

Fixed flexion deformity (FFD) is one of the common clinical entities presented to PMR outpatient department for rehabilitation. FFD of hip is often associated with fixed knee flexion and hyperextension of the lumbar spine. Most of the time flexion deformity occurs due to contracture of joint capsule or of muscle. FFD is the commonest deformity of the hip and very common in some form of arthritis like pyogenic, tuberculous, osteo-arthritis, rheumatoid arthritis etc. In India tuberculosis infection is really prevalent particularly in lower socioeconomic condition.

Case Report:

A seventeen years young lady was referred to outpatient department of Physical Medicine and Rehabilitation from department of Orthopaedics for rehabilitation of FFD of hip. This cachectic lady was suffering pain in her left lower limb for five years and difficulty in walking for last two years. She was absolutely fine 5 years back. Suddenly severe pain started in her left knee and lower part of left thigh and general practitioner treated her with analgesics and cold therapy. At that time ASO titre was done to rule out rheumatic fever although none of the other clinical features of modified Jone’s criteria were present. Then she was seen by a specialist physician who investigated her to rule out oligo-articular variety of juvenile spondyloarthropathy. All the serological markers like rheumatoid factor, antinuclear factor, HLA B27 were negative. At that time she was also suffering from little bit of left hip pain. Subsequently she has been advised to go for physiotherapy. Different types of modalities including deep heat therapy and IFT were applied without any positive
outcome. Physiotherapist assessed that pain as a radiating pain from low back. After reviewing the x-ray (Fig 1) of lumbosacral spine the therapist suggested her to take an opinion from neurologist because there was spina bifida of fifth lumbar vertebra.

But her parents brought her to an orthopaedician’s clinic. The orthopaedician diagnosed her as a case of Parthe’s disease considering hip pain of a 17-year-old girl and thought knee pain as referred pain from her hip. That’s why he advised her to go for a hip x-ray and prescribed only analgesics. Interestingly the x-ray (Fig 2) was reported as avascular necrosis of femoral head. In the meantime the pain was increasing in spite of so many consultation and little bit of deformity was also developed. The patient was then assessed by a neurologist who documented this pain as musculoskeletal problem. He sent the patient to another orthopaedic team who diagnosed her as a case of FFD and referred her to department of Physical Medicine and Rehabilitation.

Eventually the patient visited PMR OPD with FFD of hip and knee. At that time pain score was minimal without any constitutional symptoms, palpable lymphadenopathy, positive respiratory signs. She was cachectic with history of weight loss, anorexia but no vomiting. She had no diarrhoea, palpitation, tachycardia, goitre. On goniometric examination her knee range of motion (ROM) was 45-75 degree and hip ROM was 30-60 degree. There was no clearly palpable mass anywhere in her body.

Keeping in mind about hip pain, FFD, cachexia, weight loss a differential diagnoses of tuberculosis infection and neoplasia in or around the hip were considered. For confirmatory diagnoses routine blood test, chest x-ray, Mantoux test and a repeat x-ray of hip and knee were advised. As per her investigation reports she had mild anaemia and raised ESR without any lymphocytosis or lung abnormality and Mantoux positivity. Interestingly little periosteal reaction of femur was noted in her x-ray (Fig 3).
That’s why a CT scan and CT guided FNAC were advised to rule any neoplasia. In CT scan (Figs 4-6) a big mass was seen involving gluteal and thigh muscle of her left side with bit of involvement of periosteum. Even the mass was extended up to the left sphincter ani muscle. Fortunately this young girl was continent throughout the disease course. According to CT scan report there was strong possibility of soft tissue sarcoma of the left lower limb.

CT guided FNAC (Fig 7) from the mass picked up the diagnoses. After thorough review of the pathological slides (Figs 8–9) it was seen that multiple clumps of benign spindle cells having blunt nuclei without significant mitotic activity confirming histopathologically as a case of benign fibrous histiocytoma. After that the patient was sent to surgical team for wide local excision of the mass. At last patient became pain free and independent with postsurgical rehabilitation programme.

Discussion:
Benign fibrous histiocytoma is a rare entity that was first described by Dahlin in 1978. This lesion occurs most
frequently in soft tissue and less often in bones. Benign fibrous histiocytoma may occur in the diaphysis or epiphysis of long bone or in the pelvis. Common bones involved are femur, tibia, humerus etc, according to Oxford text book of Orthopaedics and Trauma. In this young girl the mass originating from the glutei and hamstring evaded femur. According to the literature it may occur in any age group though most common age of presentation is 30 to 40 years. Our patient presented at much younger age group. Although it may present in much earlier stage of life even in first year. Benign fibrous histiocytoma is radiologically seen as well-defined lytic expanding lesion with little periosteal reaction. In contrast to non-ossifying fibroma this lesion is considered as true neoplasm. Because of its tendency for local recurrence extended curettage or wide excision is recommended. Preoperative assessment of neurovascular entrapment and involvement of muscles or compartments and expected residual functional capacity of the limb after surgery are very helpful for overall prognoses of the patient.

**Conclusion:**
Although benign fibrous histiocytoma is a very rare clinical condition but may be a cause of FFD of hip and knee. Cross checking of clinical diagnoses and investigations for confirmatory diagnosis is an integral part of rehabilitation process.

**References:**