



ISSN: 0976-3376

Available Online at <http://www.journalajst.com>

ASIAN JOURNAL OF
SCIENCE AND TECHNOLOGY

Asian Journal of Science and Technology
Vol. 10, Issue, 07, pp.9889-9891, July, 2019

RESEARCH ARTICLE

MANAGEMENT OF OLLIERS DISEASE AND ITS COMPLICATIONS: 11 YEARS PROSPECTIVE STUDY OF A CASE REPORT

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ARTICLE INFO

Article History:

Received 19th April, 2019
Received in revised form
14th May, 2019
Accepted 17th June, 2019
Published online 31st July, 2019

Key words:

Enchondromas, Conservative
Treatment, Corticotomy, Limb
Lengthening,
Partial Epiphyseal Arrest.

ABSTRACT

7 yr old male child presented to us for shortening of left lower limb by 2 cm with mild pain thigh and leg before 11yrs. We followed him till today.on investigation and biopsy from tibia found multiple enchondromas even in opposit lower limb but not much significant symptomatic. 4 yrs later pt developed pathological fracture in lower end left femur treated surgically .at the onset of growth spurt pt developed limb length discrepancy and varus angular deformity at left knee.pt underwent corrective surgery in the form of partial epiphyseal growth arrest surgery and limb lengthening surgery for tibia as its contribution in shortening was more ,by rail Road fixation and corticotomy at proximal end . Though literature advocates ring fixator but we found it more convenient. at the end of 9 months we achieved our goal. 15.6 degree of varus was reduced to 6 degree and was compensated at valgus of 6 degrees. Even after 4 ½ yrs of last surgery patient is doing very well.

Citation: Kuldeep nahar, 2019. "Management of olliers disease and its complications: 11 years prospective study of a case report", *Asian Journal of Science and Technology*, 10, (07), 9889-9891.

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INTRODUCTION

Enchondromas or chondroblastoma or low grade chondral series tumours are relatively common intramedullary cartilage tumours commonly associated with olliers syndrome and Maffucci syndromes (enchondroma +skin hemangioma) .Peak incidence is in between 10-30years of age, ranging from 3-10% of all bone tumours and 12-24%of benign bone tumours. Clinically they are asymptomatic rarely painful and presented as incidental finding may present as limb length discrepancy .These are cartilage lobules surrounded by normal one or ring and arcs patterns if they are ossified. Basically they arise from rests of growth plate entrapped in metaphysis of growing bone ends.they highly resemble to low grade chondrosarcomas histologically diagnosis depends upon clinical, radiological, pathological correlation. Grossly lesions are <3cms, translucent, nodular & grossly. Grey blue these are typically located in a central or eccentric position in the medullary cavity of tubular cavities of small and long tubular bones of hand, feet, humerus, femur, tibia. Rarely they appear in pelvis, ribs, scapula. as vary rare exophytic growth pattern known as enchondroma protrubarence. Radiographically they are small lytic lesions with non aggressive features:

.narrow zone of transition
.sharply defined margins
.+|_ chondroid calcification (rings& arcs calcification)
Often no matrix mineralisation (purely lytic)
.+|_ expansile
May have mild endosteal scalloping
Should not grow through cortex
. pertinent negative No gross bone destruction, no periosteal reaction no Soft tissue mass.

Enchondromas mainly seen in metaphysis, may be in diaphysis but rarely in epiphysis as their origin from growth plate. MRI is useful in evaluating the soft tissue extension and for confirmation of diagnosis.they appear well circumscribed, somewhat lobulated replacing marrow

- T1: intermediate to low signal
- T1c+[g] (a) enhancement is variable & maybe seen both peripherally or of translesional septae (b) similar pattern may be seen in chondrosarcoma
- T2(a)typically of background intense high signal (b)they can be focal regions of signal drop out where calcification present © No bone marrow or soft tissue oedema
- Differentiation of enchondroma &low grade chondrosarcoma is problematic on Mri Nuclear medicine may show increased uptake on bone scan.intense uptake may show in pathological fracture & soft tissue expansion.

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Pre-op before biopsy



Fixation of fracture



Pre op before biopsy



Correction of varus



Pre-op pathological fracture



Limb lengthening

Majority of enchondromas remain asymptomatic and require no treatment pathological fracture are treated with curettage and bone grafting recurrence is reported in 2-15% and 25 % up to the age of 40 yrs is considered malignant. Differential diagnosis on MRI include Bone infarcts, Chondrosarcoma, Intraosseous ganglion and Other benign lytic bone lesions:-



Final follow up results

Lytic metastasis to Bone Granulomatous disease, sarcoidosis, tuberculosis.

METHODS AND MATERIALS

A 7yrs old male boy presented with pain in thigh and leg left lower limb. Rom of knee was full and no neurological deficit. xray showed multiple osteolytic lesion with sclerosis in femur and tibia in entire length (diaphysis +metaphysis).pt underwent open biopsy of tibial lesion on 12.10.2008 it was enchondroma.pt was treated conservatively.pt presented again after 4years with pathological fracture of long oblique in nature's left femur lower end was operated by intramedullary elastic nail and plate and screw.pt was again operated for removal of implants. In August 2014, pt presented with shortening of left lower limb by 3" inches and severe varus

deformity at knee.pt was operated for temporary growth arrest surgery by putting plate at lateral growth plate in left lower end femur tibia was lengthened by putting rail road fixator and corticotomy and callotaxis and desired length and correction achieved in deformity within 9 months and again implants removed.

DISCUSSION

As per literature for Ollier's disease and Maffucci syndrome. 1 Angular deformities were common; 80 per cent of the affected femora had clinically significant varus or valgus angulation in the distal part and 42 per cent of the affected tibiae had proximal or distal deformity. The apex of the angulation, when present, was metaphyseal, with the concavity on the side that was more extensively involved by the enchondromas. Deformity in the distal part of the femur frequently required repeat osteotomy to achieve a straight bone at skeletal maturity, The extent of shortening, which always was present in the involved limb, closely paralleled the extent of involvement.

The discrepancies in limb lengths prior to surgical treatment averaged 9.8 centimeters (range, 4.3 to 35.7 centimeters). In our case it was 2 cm at time of first consultation and 8cm prior corrective surgery. (1) Epiphyseal arrest, when appropriately timed, was effective in correcting or limiting the discrepancies, but partial (medial or lateral) epiphyseal arrest to correct angular deformity was ineffective but in our case partial epiphyseal arrest of lateral femoral condyle worked very well. Some author in literature found early maturation of callous while bone lengthening procedure but we did not encounter such phenomenon.

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