

## Case Report

# Management of Rare Infantile Myofibromatosis of the Tibia by the Ilizarov External Fixator

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Submitted: 17 April 2015

Accepted: 15 August 2015

Published: 16 August 2015

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**Abstract**

Infantile myofibromatosis is considered as the most common fibrous tumor of infancy. Skeletal affection was reported in numerous articles. In this study we have treated two cases with severe tibial deformities using the Ilizarov frame. The Ilizarov was found useful in correction of the deformity as well as in limb length discrepancy management. Recurrence however, took place in one case and the resultant deformity was treated adequately using the Ilizarov frame. It was concluded that the Ilizarov frame was adequate for management of infantile myofibromatosis of the tibia for deformity correction and lengthening.

**INTRODUCTION**

Infantile myofibromatosis is a rare mesenchymal disorder of early childhood characterized by the formation of tumors in skin, muscle, viscera, bone, and subcutaneous tissue [1-3]. It may present with a solitary form or a multi-centric form. The condition was previously known as congenital generalized myofibromatosis. It affects almost exclusively infants and young children. It was felt to be a rarely seen disorder that was frequently fatal [2]. However, a relatively recent summary of 155 affected infants found that most had been reported since 1980 [3]. Since this publication we have cared for two children with this disorder, and more infants were found in the literature since [4-6].

This disorder is considered as the most common fibrous tumor of infancy, and may mimic numerous other clinical entities. [1,7] taking in consideration that this condition is rare and has been frequently mis-diagnosed, we present our two cases, with the clinical manifestations, histopathologic features, prognosis, and management of the resultant deformity of the tibia by the Ilizarov frame.

**PATIENTS AND METHODS**

Between October 1990, and October 2009, we reported two cases of infantile myofibromatosis (IM), the age of the patients at time of presentation was; 4 and 5 years.

**Case 1**

A male child 4 years, presented with an insidious onset and

a slow progressive course that ended in 11 cm of shortening of the left leg. There was an antero-medial angulation of 90° at the junction of the middle and distal thirds of the tibia [Figure 1]. Violet discoloration of the skin overlying the apex of the deformity was noted. The left foot was medially rotated to about 180°, with the heel facing forwards and the dorsum of the forefoot facing backwards. Plain X-ray of the left lower limb was performed in Antero-posterior view and lateral view [Figure 2]. A CT scan was performed, to measure the limb length discrepancy [LLD], and a CT scan of the thorax was performed to exclude other visceral lesions.

An open biopsy was taken and revealed; I. Gross Picture; A, 2 pieces of a core, white soft tissue, the first one measuring 2x 0.1 cm. the second measures 0.5x 0.1 cm, submitted in toto.

B, 2 pieces of a white linear tissue, measuring 2x 0.5x0.1 cm, submitted in toto.

The microscopic picture revealed; A&B sections show a tumor composed of an admixture of fibroblast like spindle cells and plump fusion cells resembling smooth muscle fibers arranged in short bundles and associated with considerable amount of collagen focally. Foci of hemangiopericytoma like areas are seen as well as fragments of coagulative necrosis. No pleomorphism or mitotic figures were seen. The diagnosis was consistent of infantile myofibromatosis [IM].

The deformity was treated by an Ilizarov frame [Figure 3,4], a corticotomy site at the proximal tibial segment was done and gradual distraction and correction was applied. Antero-medial angulation and rotation of the tibia were corrected simultaneously

in the frame [Figure 5]. The derotation construct of the frame was useful and gradual derotation was performed, until the foot was put in the proper plane and regained its normal position. After correction, and consolidation of the regenerate, the frame was removed [Figure 6], and a full time, day and night, brace was applied. LLD was corrected and the foot was plantigrade by the end of the lengthening procedure.

Unfortunately, after 3 years of follow up, recurrence of the deformity took place. This was again corrected with the use of the Ilizarov frame, and re-lengthening was simultaneously performed to equalize the LLD. The patients are still under follow-up.

## Case 2

Another male patient aged 5 years at presentation, similar to



Figure 1 Clinical appearance of the case at presentation.



Figure 2 Plain X ray of the affected leg, showing the osteolytic lesion.

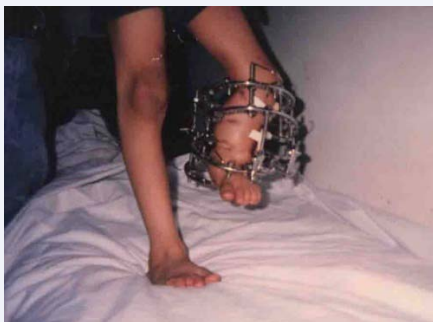


Figure 3 Clinical photo of the patient during lengthening by the Ilizarov frame.

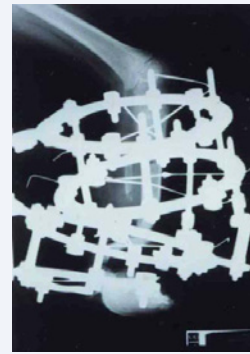


Figure 4 X ray Lateral view after application of the Frame.

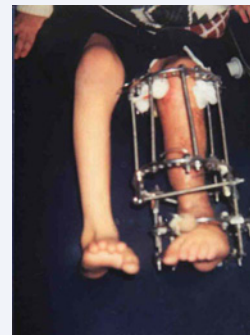


Figure 5 Clinical appearance after equalization of the LLD.



Figure 6 Clinical appearance after frame removal.

case number 1, there was a lesion at the lower third of the leg, with a LLD of about 8 cm, the foot was angulated and rotated but to a lesser extent. After thorough examination and investigations, including open biopsy revealed a lytic radiolucent lesion, and the diagnosis was made as IM. Corrective surgery, by en bloc resection of the osteolytic nodule, a corticotomy at the proximal tibia, and mounting of an Ilizarov frame was performed. The foot and ankle were freely mobile at presentation, and a full weight-bearing plantigrade foot was achieved by the end of the lengthening procedure. After frame removal, the brace was worn in a full time regime, and the patient is followed up till present, with no signs or recurrence.

## DISCUSSION

IM has been a known entity since 1954 [8]. It represents the

most common fibrous tumor of infancy. [1-3] Various types were described with variable prognosis and behavior [1]. The extra-skeletal lesions present on X-ray examination as a soft tissue mass, often with small foci of calcification. Skeletal affection usually presents with variable size radiolucent lytic lesions often described as, cystic defects. Some of the lesions may present with an eccentric growth or as a circumscribed lytic lesion with sclerotic borders [1-3]. The skull, femur, tibia, spine, and ribs, were the most commonly involved bones.

Microscopically, the solitary and multiple lesions varied little in their appearance. All were composed of short, curving bundles of fusiform, collagen-producing cells, which showed staining characteristics intermediate between fibroblasts and smooth muscle cells. The cells formed distinct nodules, often with evidence of necrosis (with or without calcification), or a hemangiopericytoma-like cellular pattern in their center. The nodules are for the most part well circumscribed, but occasionally short bundles of spindle cells extended in a finger like fashion into the neighboring tissues [9].

Differential diagnosis of IM is dependent upon the location and number of nodules at presentation. Typical bone lesions may resemble those seen in fibrous dysplasia, neurofibromatosis, metastatic neuroblastoma, lymphangiomatosis, Leterer-Siwe disease, eosinophilic granuloma, and familial multiple osteogenic neurofibromata. Presentation of soft tissue lesions with conjunction with skeletal lesion aids in narrowing the diagnosis if IM is considered along with the possibility of metastatic disease. Diagnosis of IM ultimately depends on adequate tissue sampling. This in an important step which may obviate the need for performing aggressive treatment, particularly in cases of multiple lesions without visceral involvement, where spontaneous regression of the lesions without treatment is the general rule. Appropriate radiographic evaluation in children diagnosed with IM, should include, chest X ray and CT, skeletal survey, and evaluation of the gastrointestinal tract and the cardiovascular system to search for all identifiable lesions [10-12].

Bony deformities, as well as limb length discrepancy was reported in this study due to affection of the diaphyseal part of the lower limb bones. The Ilizarov frame was found very useful in the course of management of these rare disorders, to correct

the resultant deformity as well as equalization of the LLD. Although this was a good tool of treatment and full correction of the deformity was possible, recurrence occurred and is to be anticipated.

The main shortcoming of this study is that it is a retrospective study, and of course the few number of the included patients due to the rarity of this condition.

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### Cite this article

El-Sayed M, Hosny G, Rizk O (2015) Management of Rare Infantile Myofibromatosis of the Tibia by the Ilizarov External Fixator. *Ann Orthop Rheumatol* 3(3): 1053.