Myositis Ossificans Progressiva

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Citation

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Abstract

Myositis ossificans progressiva (MOP) or fibrodysplasia ossificans progressiva is a severe extremely rare condition of ectopic ossification with primary involvement of the skeletal muscles associated with charecteristic skeletal abnormalities with an average incidence of 1/10million. It was first described by Guy Paten in the year 1962.

INTRODUCTION

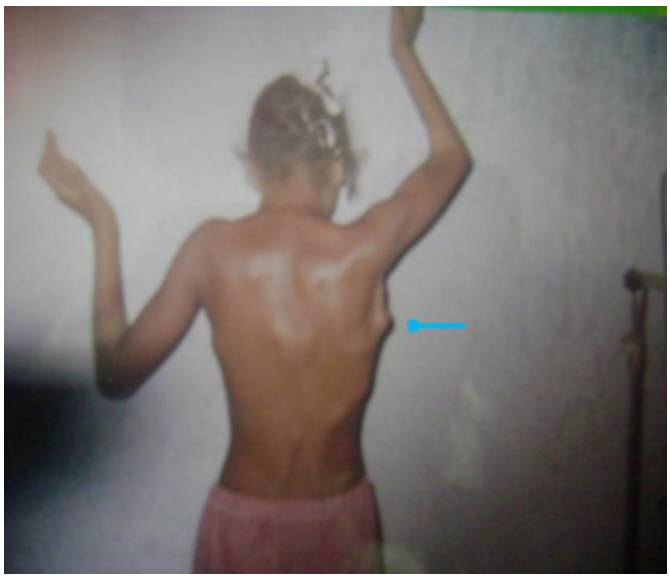
MOP or FOP is a rare autosomal dominant collagen disorder with complete penetrance and variable expressivity characterised by skeletal deformity & progressive heterotopic osteogenesis. Shore et al reported mapping FOP to chromosome 2q23-24 by linkage analysis. They also identified an identical heterozygous mutation (617G R206H) in the glycine-serine (GS) domain of the activin A receptor type I (ACVR1) gene, a bone morphogenic protein (BMP) type I receptor in all affected individuals and are thought to be plausible candidate genes. There is no predilection for any sex and It occurs with equal prevalence in all the races. Average age of onset is 5yrs, with reported onset ranges from fetus to 25yrs.

CASE REPORT

A 10 years old girl presented to us with chief complaints of multiple bony hard swellings involving both upper and lower back for 2 years, pain right shoulder for 1 year and neck pain for 6 months.

Figure 1

Figure 1



2 yrs back, she started with swelling over right scapular region which gradually progressed distally to right ribcage. Swelling was hard in consistency, which increased in size gradually. The swelling was associated with pain right shoulder. This was followed by appearance of similar swellings over left lower scapular region and right lumbar region. 6 months back she started with pain neck and painful neck movements. On examination, she had multiple swellings involving upper & lower back, right ribcage, right side of the neck and hallux valgus right foot. The largest swelling is present over right ribcage below the lateral border of scapula. On palpation bony hard cord like structures are palpable on right side of the ribcage, right lumbar region, left & right scapular region, left & right side of the neck . The overlying skin is free, however the bony

bands were adherent to the underlying bone. Overhead abduction of right shoulder was restricted.

Figure 2

Figure 2

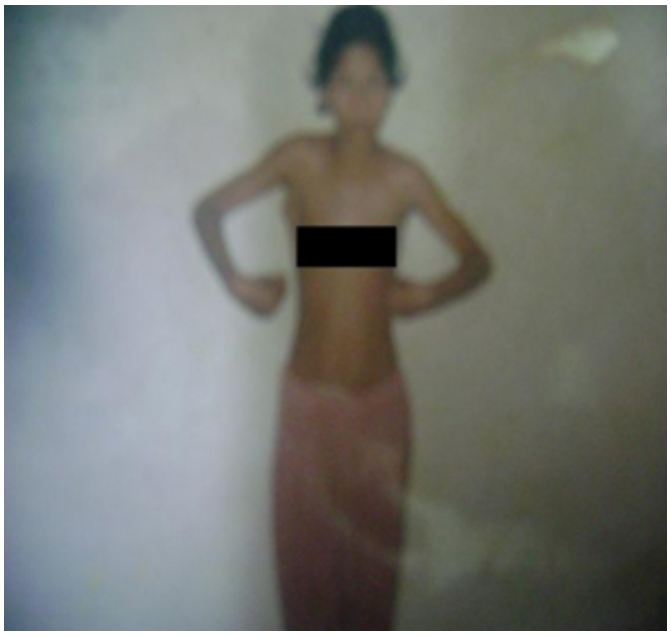
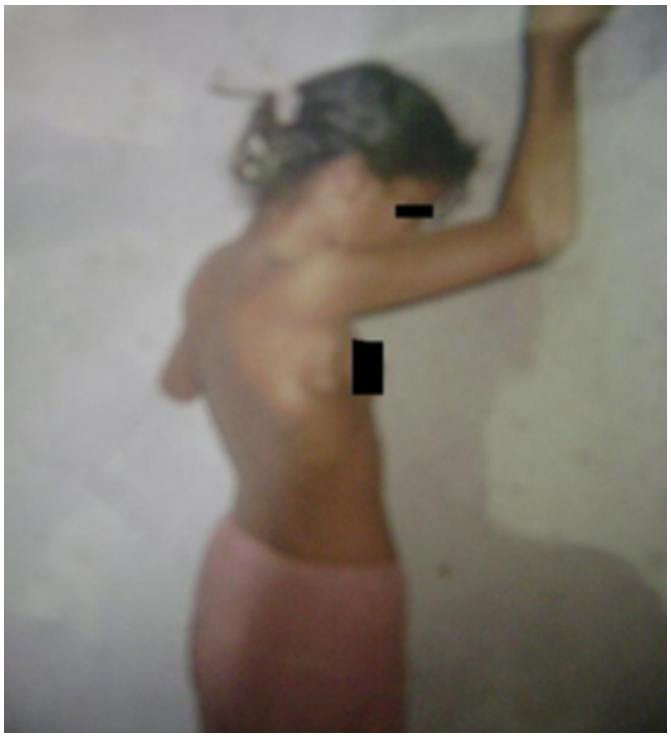


Figure 3

Figure 3



Blood biochemistry: Serum proteins, albumin or A:G ratio is within normal limits. There is no abnormality in serum calcium or phosphorous level. ESR is raised to 125, as occurs during an acute episode.

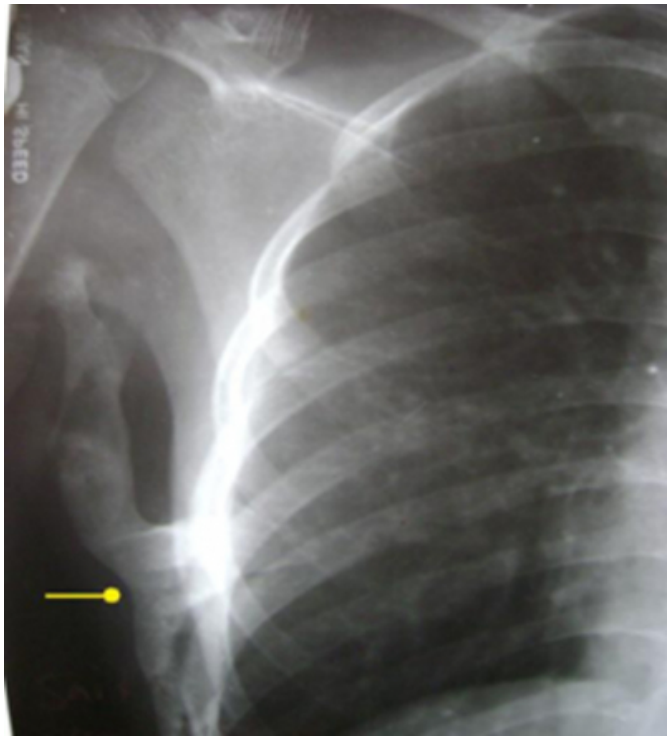
RADIOLOGICAL FEATURES

The roentgenographic findings show radio-opaque shadows, due the heterotopic bone formation, in the various regions of the body.

X-ray right shoulder AP view shows, cord like radio-opaque lesion in the soft tissue plane over the lateral aspect of the Rt. scapula going downwards & involving right ribcage and going further downwards to involve the soft tissue.

Figure 4

Figure 4



X-ray of the dorsal spine shows a radio-opaque band of ossification with involvement of surrounding soft tissue.

Figure 5

Figure 5

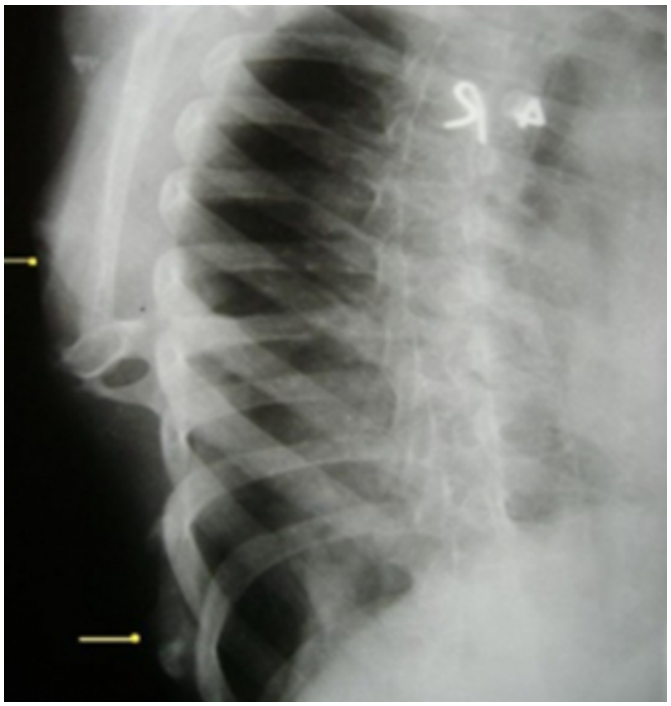
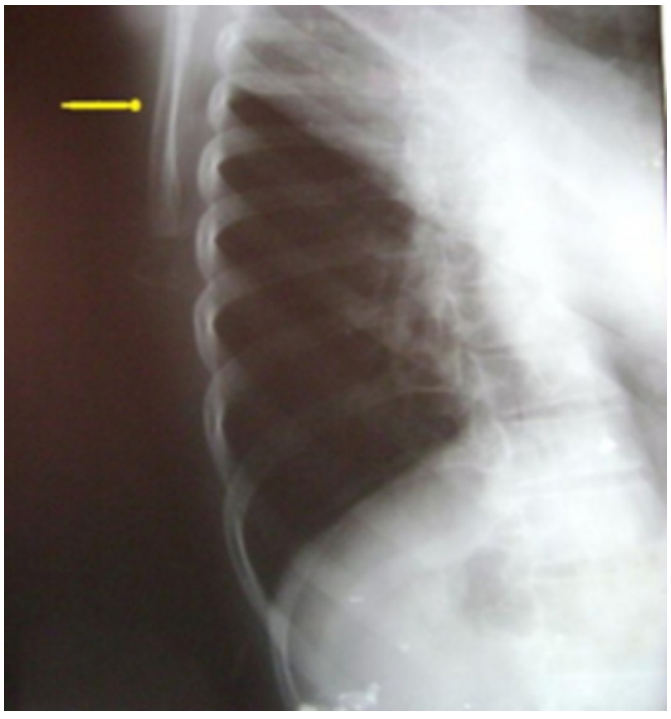


Figure 7

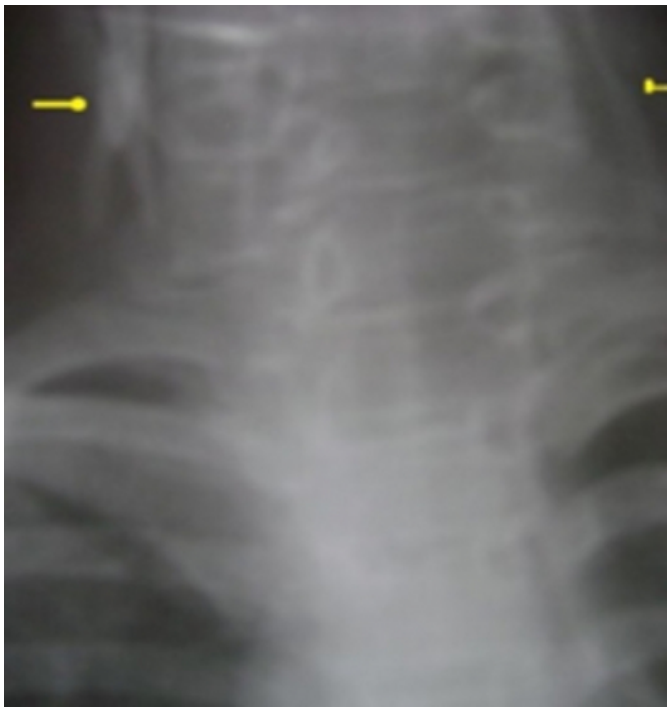
Figure 6



X-ray of cervical spine shows vertical cord like radio-opaque shadows on either side of the spine, Suggestive of heterotropic ossification of paravertebral muscles.

Figure 8

Figure 7



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CT- scan: As the radio-opaque cord like lesions of heterotropic ossification were evident on radiographs and cost was the other factor and it would not help the pt. in any ways so we did not perform it.

Histopathology: As it precipitates new lesions and therefore is contraindicated in the MOP, so we didn't perform.

TREATMENT

After extensive research through various texts and internet we couldn't find any treatment for this lethal disease. We managed the patient symptomatically.

DISCUSSION

MOP / FOP is the rare autosomal dominant connective tissue disorder that results in the heterotopic osteogenesis & skeletal deformity with no definitive treatment till date. There is little convincing evidence that any form of treatment alters the progress of the disease in myositis ossificans progressiva. Treatments that have been used include a diet reduced in vitamin D and calcium, the avoidance of sunshine, and treatment with corticosteroids. Other treatment strategies include beryllium, vitamins B and E and penicillamine. Drugs that block calcification have also been used, including EDTA (disodium ethylene diamine tetraacetic acid), isotretinoin and etidronate, without proven

benefit. There is one report of treatment with ascorbic acid. Surgical resection of the ossified sites is usually unsuccessful, with recurrence at the same site.

Search is still on regarding the proper treatment modality of myositis ossificans progressive, Let us hope in the coming era a treatment plan is figured out for this dreaded disease.

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