ARC Journal of Clinical Case Reports
Volume 10, Issue 3, 2024, PP 6-9
ISSN No. (Online) 2455-9806
DOI: https://doi.org/10.20431/2455-9806.1003002
www.arcjournals.org

A Case of Myositis Ossificans Progressiva from Sudan

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Abstract

Myositis ossificans progressiva is a one diagnosis that not usually seen on daily basis in the emergency rooms, or even in referred clinics. In fact, specialty training could pass without encountering a case. The unique etiology, extensive deformity and presentation, and the management protocol encouraged the authors to present this case of a Sudanese adolescent female who presented to the refereed clinic in Port Sudan hospital in eastern Sudan.

Keywords: *Myositis ossificans, deformity, ankylosis, autosomal dominant.*

1. Introduction

Myositis ossificans progressiva (MOP), also known as fibroplasia ossificans progressiva, is a rare autosomal dominant disease where less than 1000 cases of have been reported globally. It has varying degrees of predominance, however, sporadic cases has also been reported[i].

The disease was first identified in the seventeenth century, and till now there is no clear pathophysiology [ii], and the most involved area is the posterior cervical area in the neck[iii]. Inflammatory process occurs leading to edema and then calcification and affected mobility.

The current treatment modality comprises of steroids, bisphosphonate, and there is a role of ascorbic acid in modulating the synthesis of procollagen III[iv].

Here we report a case of MOP, presentation and clinical manifestation in a young female from Sudan, as well as treatment options.

2. Case Report

12 years old female presented to the pediatric hospital in Port Sudan city with hard masses in several areas of her body and deformities. It started since early infancy, but had worsened over time.

By the age of 2 years she complained of pain and movement difficulties, short digits and atrophied fingers. On examination, there were multiple contractures mostly in her left upper and lower limbs as well as her spine. Her posture is scoliotic with high degree of lumbar lordosis, and elbow limitation. She also has cervical and dorso-lumbar stiffness. Patient has bony prominence in her left posterior chest wall, right, and left shoulder. Her left hip is stiff in extension position. The patient has a stiff anterior thigh muscles (iliacus, psoas major, pectineus, rectus femoris, adductor longus, Sartorius, vastus latralis, vastus medialis). She has also severe left sided elbow limitation. Her gait is equine with left side knee extension. There was stiffness in her wrist in a downward position, and there is shortened foot phalanx (Figure1A-F).

Figure: *Skeletal abnormalities detected in the patient. Scoliosis (1A)*

Stiff left wrist on extension (1B)

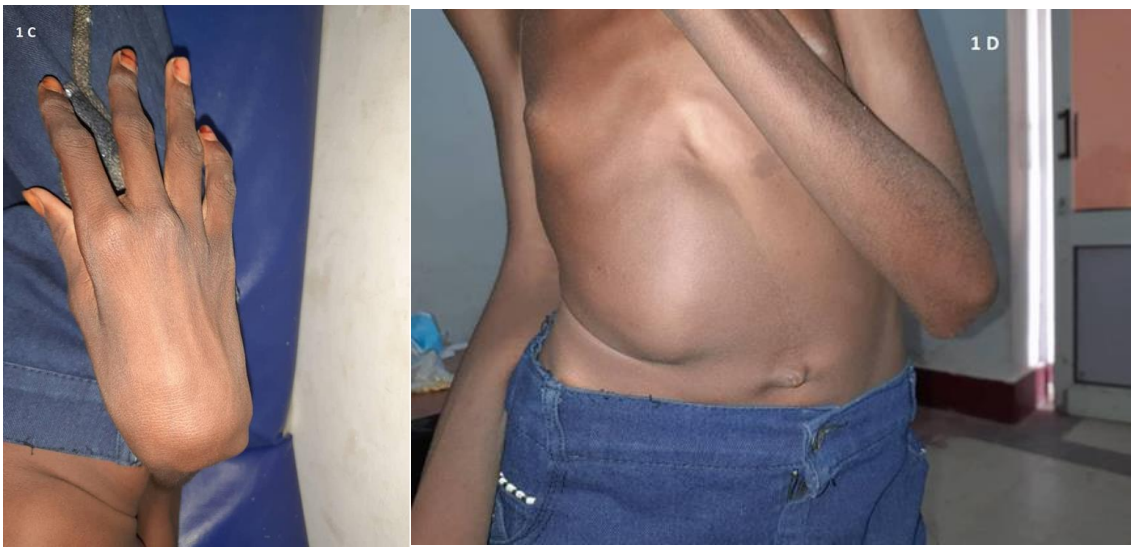
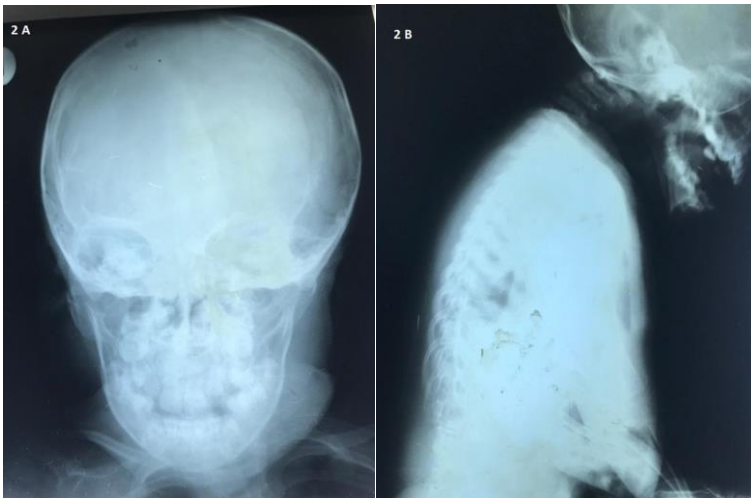


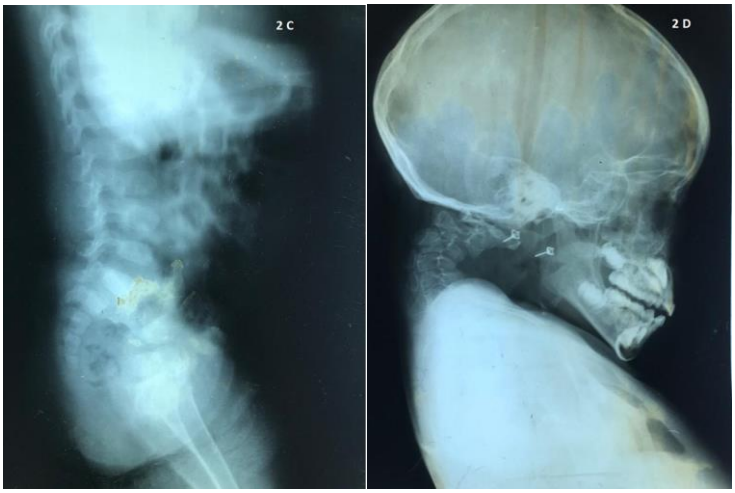
Figure: *Stiff right wrist on flexion (1C); Stiff left elbow on flexion (1D)*

Figure: *Prominent nodule on left shoulder inferior to the clavicle (1E); and short phalanges of the foot (1F).*

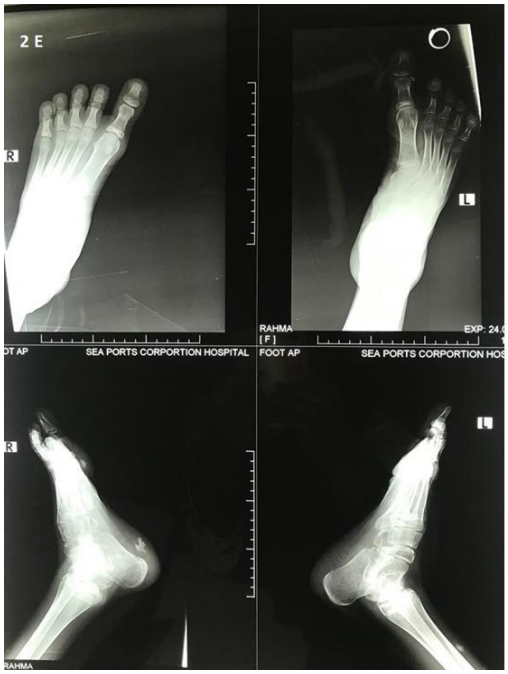
X-rays showed hyper-calcified teeth, ankle, wrist, right hip, and metatarsophalangeal area. Dense mesh of cocoon-like webs are seen in the thoracic lateral views (Figure 2A-F).



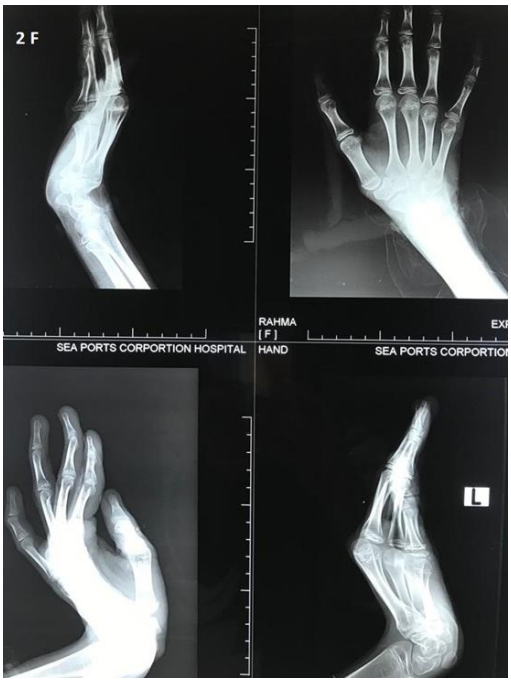
Radiological findings in the patient. Hyper ossified skull (2A); cocoon mesh ossification of soft tissue in lateral thoraco-abdominal view (2B);



Hyper-dense ossification of the right hip (2C); dense ossified soft tissue and teeth (2D)



Dense ossified ankles and metatarsus (2E)



Dense bone structure on left wrist and distal radius and ulna (2F).

Laboratory tests for calcium, phosphorus, alkaline phosphatase, urea, creatinine and parathyroid hormone were found to have normal values. Patient received steroids-in the form of prednisolone, and vitamin C. The patient was discharged in a stable condition. The patient improved after management, lesser pain and stiffness.

In conclusion, MOP is a rare disorder with uncertain pathogenesis and possible inheritance patter. This case presented with extensive ossifications in the trunk and limbs. Conservative management stabilized the condition and improved her lifestyle.

DECLARATION

Conflict of Interest statement: All authors have no conflicts to declare.

Funding: none.

Ethical Approval: Obtained from the hospital administration.

Consent: Verbal informed consent was obtained from the patient and her family for publication of this case report with accompanying images.

Guarantor: Osama Abbadi.

Authorship: Conseptualization: AAM; Methodology: LSH; Investigation: AH; Formal analysis: OSA; Data curation: LSH; Visualization: AAM; Writing original draft: LSH and OSA; Writing-review: AH

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Citation: *Amel Aziz Malik et al. A Case of Myositis Ossificans Progressiva from Sudan. ARC Journal of Clinical Case Reports. 2024; 10(3):6-9. DOI: http://dx.doi.org/10.20431/2455-9806.1003002.*