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Myositis Ossificans Progressiva: a rare case report

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Abstract: *Myositis ossificans progressiva is a rare congenital connective tissue disease characterized by wide spread soft tissue ossification and congenital stigmata of the musculoskeletal system. We report here about one female child 11-year old with bilateral hypoplastic hallux and little finger, multiple nodules over back, restricted chest expansion, and progressive stiffness of neck, trunk and extremities that limit her activities of daily life. The purpose of reporting this case is to raise awareness about this rare entity. It highlights the importance of early diagnosis as the key part of management to reduce the propensity of disease progression by trauma and other stimulating factors as no specific treatment is available till date.*

Keywords: *Myositis ossificans progressiva, fibrodysplasia ossificans progressiva, heterotropic ossification, autosomal dominant, bone morphogenic protein*.

I. Introduction

Myositis ossificans progressiva (MOP) otherwise known as fibrodysplasia ossificans progressiva (FOP) or Munchmeyer disease is a rare congenital connective tissue disease with less than 1000 cases reported worldwide and very few cases reported in Indian literature so far.[1] It is characterized by progressive heterotropic ossification of striated muscles, ligaments, tendons, fascia and subcutaneous tissue leading to crippling deformity and death. MOP is an autosomal dominant condition with variable penetrance.[2] It has been suggested that FOP can be caused by over production of bone morphogenic protein4 (BMP4).[3] It was described first by Patin.[4] Repeated minor trauma or injection may trigger this process.

The skeletal abnormalities include monophalangeic great toe, short first metacarpal bones, microdactyly, cleinodactyly of little finger, and abnormalities of spine. Heterotropic ossification characteristically involve the muscle of the head-neck and back in early childhood followed by the shoulders and arms. The muscles around the hips are involved later. There is characteristic sparing of muscles of facial expression, the diaphragm, laryngeal muscles, the tongue and the small muscles of the hands and feet. Common cause of death in MOP is respiratory complication.

II. Case Report

A child of 11-year old presented with difficulties in daily activities like self feeding, combing, clothing, etc. Moreover she complains of progressive stiffness all over the body interfering with her movements. Parents reported that progressive difficulties in the activities of daily life started at the age of 3 years with multiple prominent nodules over the back (Fig 1). She could perform all the necessary activities initially, but now she is unable to do anymore. She has no other associated congenital anomalies. Parents gave negative history about any trauma, injection or surgery. One of her close relative died of same condition a few years back.

On examination, the patient had a flexed stiff attitude of neck with prominent sternocleidomastoids (Fig 2), flexed and adducted attitude of both upper limbs with restriction of movement in shoulder and elbow joints, and impaired mouth opening due to restricted movement of tempero-mandibular joint. She had multiple nodules over the back, kyphotic deformity of spine with limited chest expansion of about 2-3 cm, and small little fingers (Fig 3) and great toes (Fig 4) bilaterally.

The hematological and biochemical investigation like complete hemogram, sugar, creatinine, serum calcium, phosphorus, alkaline phosphatase, CPK, ALT, AST, and LDH are within normal limit. Scleroderma antibody, Jo-1 antibody and anti-nuclear antibody (ANA) were reported negative. Audiometric report revealed normal hearing bilaterally. The X-rays showed fusion of posterior elements of cervical spine (Fig 5), bilateral bony bars extend from chest wall to humerus (Fig 6), small monophalangeic great toe with hallux valgus deformity (Fig 7), short phalanges of little fingers bilaterally (Fig 8).

III. Discussion

MOP is a rare, progressive crippling disorder with an incidence less than 1 in 10 million population. This is a mesodermal disorder with defect in reparative process causing heterotopic ossification which usually begins in early years of life.[5,6] The condition has a male preponderance. The pathogenesis of MOP remains incompletely understood.[7] The soft tissues become swollen due to edema. Back of the neck is the region most

frequently involved.[8] This edema from inflammatory process gradually becomes calcified and restricts mobility in the affected region.

There is no effective treatment so far. Numerous drugs such as bisphosphonate, squalamine, thalidomide, COX-2 inhibitors, isotretinoin, BMP4-antagonists, and steroids for acute flare up have been tried for the primary disease with limited success. Gentle guarded physiotherapy is helpful in preventing joint deformities thus improving the quality of life. Unnecessary biopsies, surgery and injections should be avoided, as they trigger or accelerate the inflammatory process, a phase that precedes ectopic calcifications.

MOP can be diagnosed by radiograph two to four weeks after the process starts. The calcification starts form periphery and progresses towards the center. The central radiolucent area is surrounded by peripheral dense opacity.[4] This pattern of calcification is unique for MOP and differentiates it from tumor where calcification starts from central part.

IV. Conclusion

A good history, clinical examination and radiological studies are important to arrive at the diagnosis of myositis ossificans progressiva. As no effective treatment has been registered yet, early diagnosis remain the main stay of management to reduce the progress of disease by trauma and other stimulating factors.

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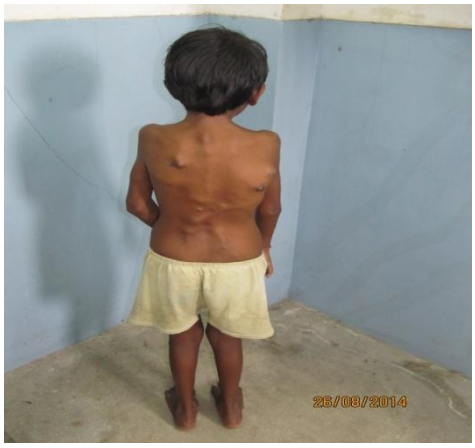

Fig 1: Multiple nodules over the back

Fig 2: Prominent sternocleidomastoids

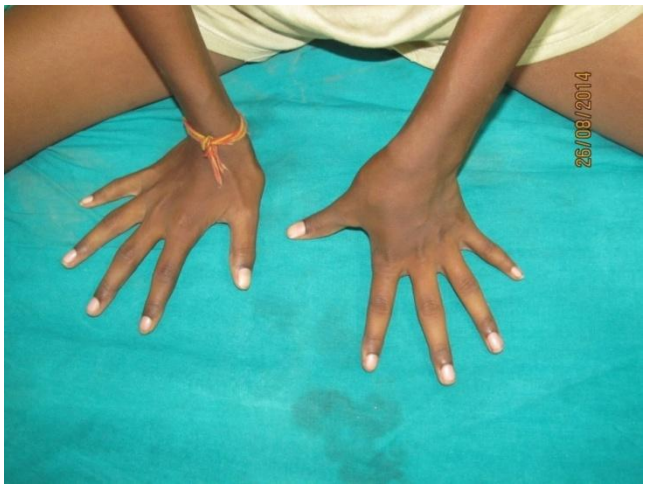

Fig 3: Small little finger of both hands

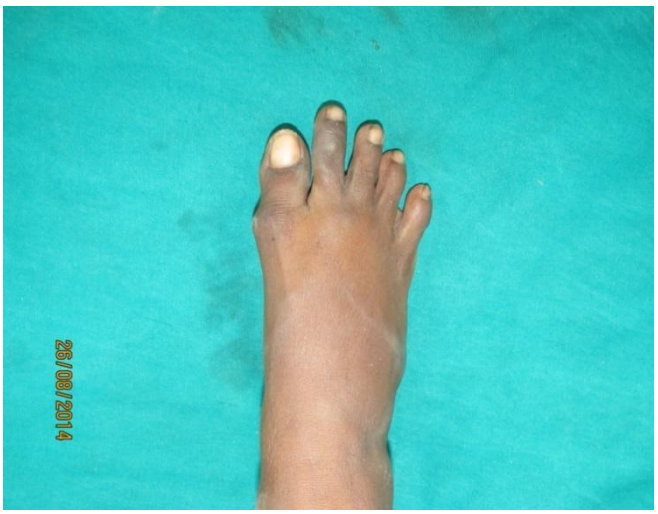

Fig 4: Small great toe with hallux valgus deformity

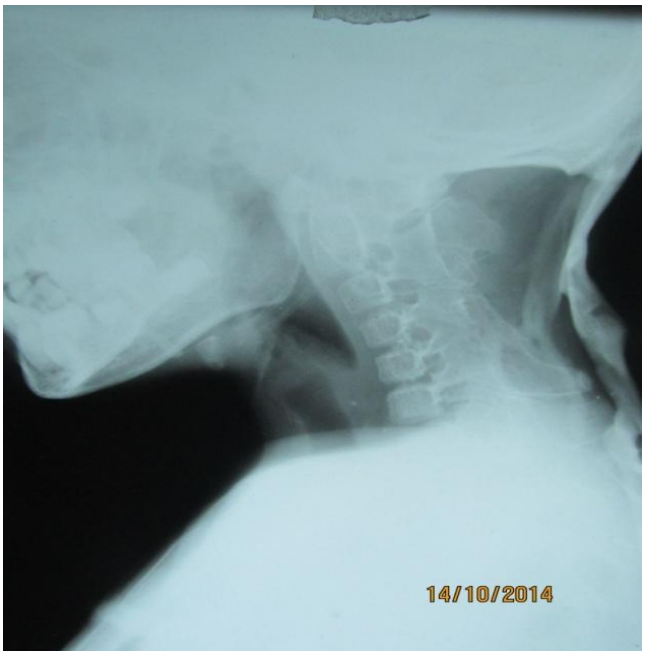

Fig 5: Fusion of posterior elements of cervical spine

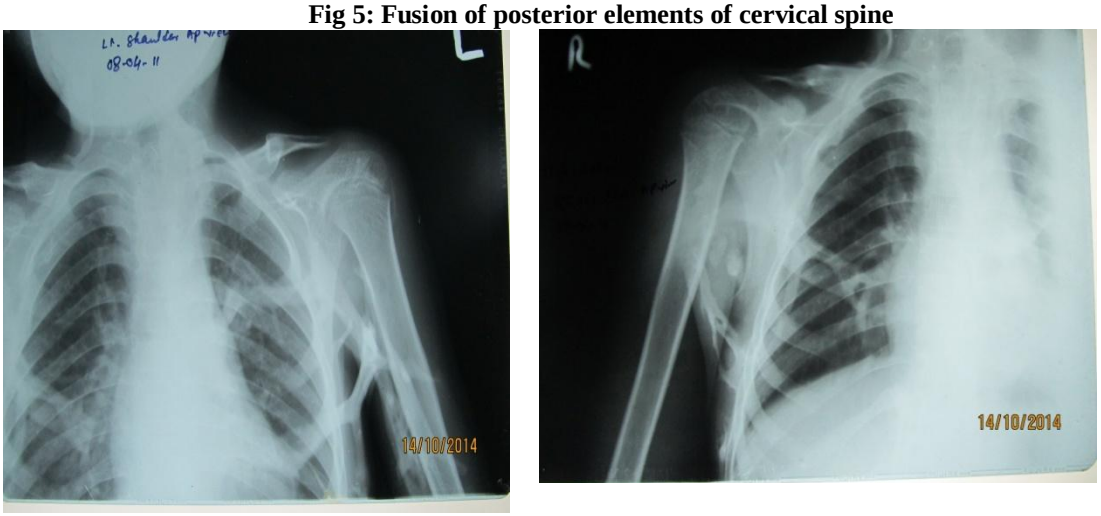

Fig 6: Bilateral bony bar between chest wall and humerus

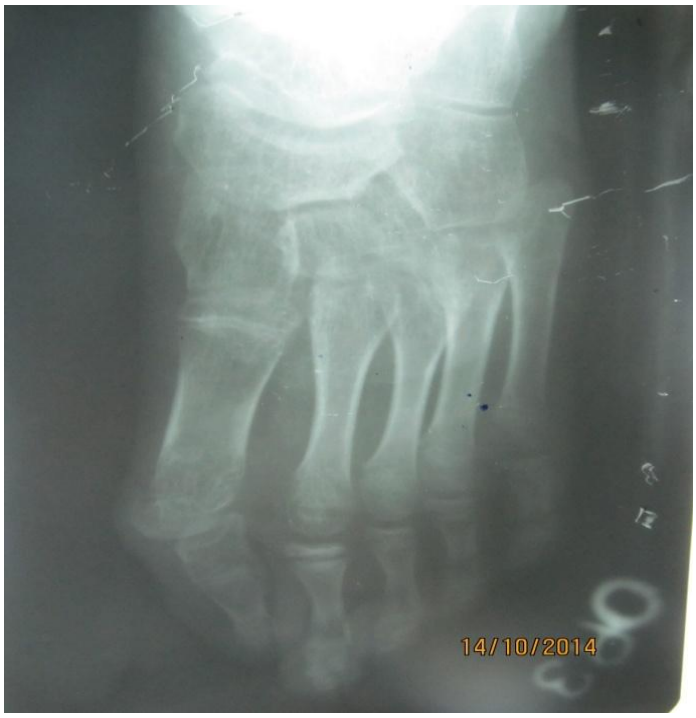

Fig 7: Monophalangeal great toe with hallux valgus deformity

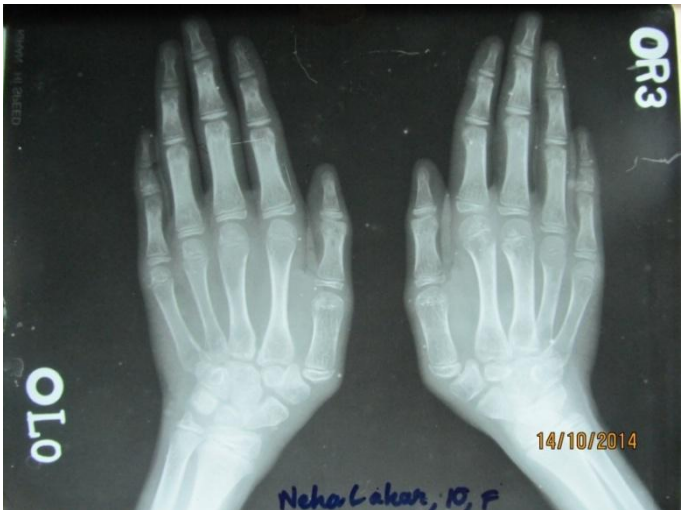

Fig 8: Bilateral short phalanges of little finger.