MYOSITIS OSSIFICANS PROGRESSIVA –A CASE REPORT

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ABSTRACT

Myositis ossificans progressiva is a rare autosomal dominant hereditary disorder, also known as Fibrodysplasia ossificans progressive (FOP) or Munchmeyer disease. Very few cases of this disorder reported worldwide. We are presenting a case of 15 year female with complaints of progressive stiffness, scoliotic deformity with restriction of movements in shoulder and elbow joint and heterotropic ossification at various sites. Early diagnosis of this disorder can be made by clinical history, examination and by imaging.

KEY-WORDS: Myositis ossificans progressiva, Hereditory disorder, Autosomal

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INTRODUCTION

Myositis ossificans progressive (MOP) is a rare hereditary disorder with autosomal dominant inheritance.[1] In most of the patient's family history is not present and it is assumed that this disorder occurs due to sporadic mutation.[2,3] MOP also knew as Fibrodysplasia ossificans progressive (FOP) reported worldwide, but in India very few cases are reported.[4] In this disorder, there is progressive heterotropic ossification of striated muscle, ligaments, tendons, and subcutaneous tissue leading to crippling deformity. The most common cause of death in these patients are respiratory complications. The responsible cause for this disorder is overproduction of bone morphogenic protein (BMP 4).[5]

Monophalangic great toe, short metacarpal bones, microdactyly, clinodactyly of a little finger, and abnormalities of the spine are found in this disorder commonly. Heterotropic ossification usually starts with muscles of head and neck and back resulting in deformity of the shoulder and spine. The muscles of hip involve late with relative sparing of the muscle of facial expression, the diaphragm, laryngeal muscle, tongue and small muscles of hand and feet.

CASE REPORT

We are presenting a case of 15 year girl who came to us with complaints of fever with flexion deformity of right lower limb. She was alright up to 10-12 year of age, then she developed progressive stiffness initially in the cervical region later on her right upper limb also involved and caused restriction of movements in her shoulder and elbow joint. She had neither any congenital abnormality nor she had a history of any trauma or surgery. On examination, she had a scoliotic posture with dorsolumber and cervical stiffness with flexed attitude of a right lower limb.

All investigations were within the normal limit. The urine microscopic examination was showing some pus cells. The X-rays show a fusion of posterior elements of the cervical spine (FIGURE-2), bilateral bony bars extends from the chest wall to the humerus (FIGURE-3), monophalangic great toe with Hallux valgus deformity (FIGURE-6), exostosis at the knee joint (FIGURE-1) and synostosis at distal radioulnar joint (FIGURE-5). On ultrasound, hypoechoic collection noted in the right psoas muscle posterior to the right kidney with raised echogenicity of the right kidney. On CECT scan, a heterogeneous collection noted in right psoas, quadratus lumborum and illiacus muscle with minimal free fluid noted in peritoneum cavity. Right kidney show striated nephrogram sign (FIGURE-4). Soft tissue ossification was seen in right psoas muscle and thigh. Multiple bony projections in the posterior aspect of the spine, anterior aspect of ribs and around the elbow with scoliotic deformity of the spine.

DISCUSSION

MOP is a rare hereditary disorder with a defect in the reparative process causing heterotropic ossification which usually begins in the early year of life.[6,7] This condition has male predominance. The soft tissue becomes swollen due to edema from an inflammatory process and becomes calcified and restrict the mobility of the affected region. The calcification starts from the periphery and progress toward the center.[7] This pattern of calcification is characteristic for the myositis ossificans progressive, while in cancer calcification starts from the center. There is no effective treatment for this disorder. Physiotherapy is helpful for joint deformity and improving the quality of life of the patient. Biopsies, surgeries, and injections should be avoided, as they start the inflammatory process, which is responsible for ectopic calcification.

CONCLUSION

Myositis ossificans progressive can be diagnosed at an early stage by history, clinical and radiological examination and it is important to start the early and proper management of the disease. Biopsies, surgeries, and injections should be avoided, as they can aggravate the disease. However, physiotherapy is helpful to prevent the deformities and improving the quality of life of the patient.

Conflicts of Interest: none.
Source of Funding: Nil.

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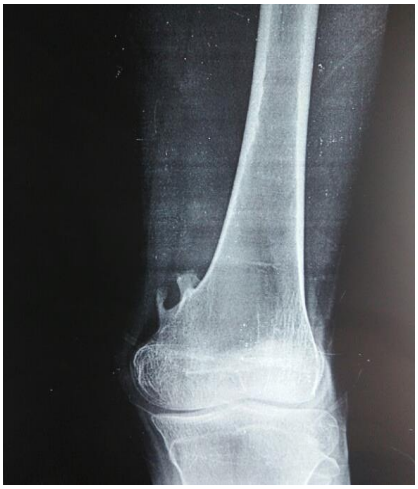

Figure-1: Xray knee joint showing multiple exostosis around knee joint.

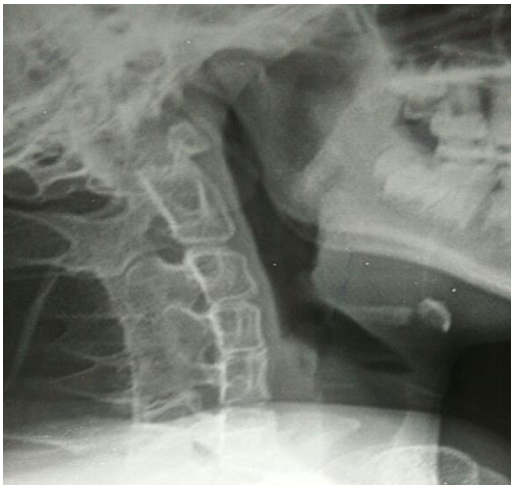

Figure-2: Xray cervical spine showing fusion of posterior elements of cervical spine.

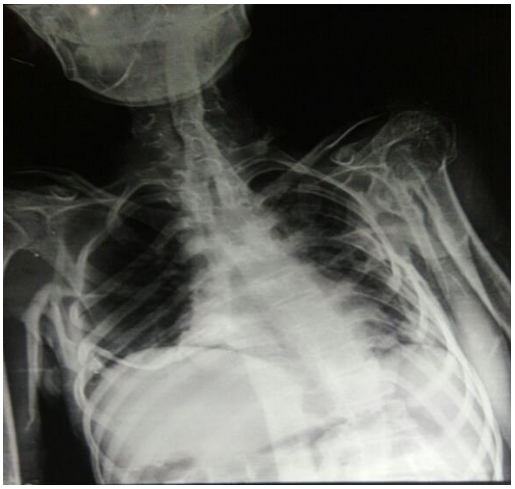

Figure-3: Chest xray showing scoliotic posture along with heretropic ossification noted along chest wall on right side.

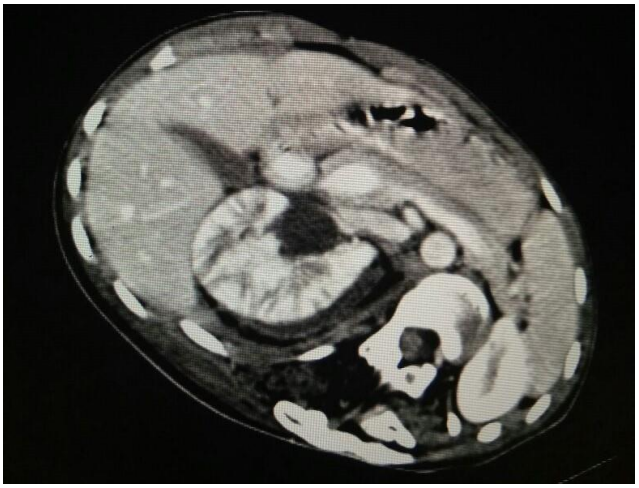

Figure-4: CECT scan showing hypodens collectionin right psoas muscle with striated nephrogram in right kidey

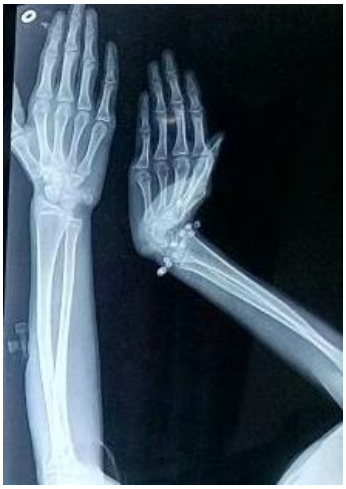

Figure-5: Xray forearm with wrist joint showing synostosis at distal radio ulnar joint.

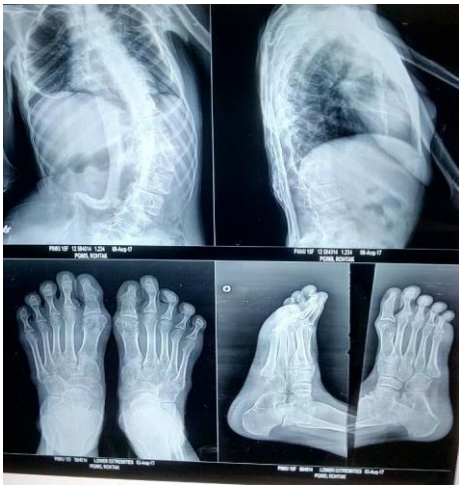

Figure-6: Chest X-ray showing scoliotic posture with bony bar and xray foot showing monophalangic great toess.