

# Myositis Ossificans Progressiva in the Whole Spine: A Case Report

Ebrahim Ghayem Hasankhani<sup>1</sup>, Farzad Omid-Kashani<sup>1</sup>, Seyed Reza Habibzadeh Shojaie<sup>1</sup>, and Golnaz Ghayem Hasankhani<sup>2</sup>

<sup>1</sup> Department of Orthopedics and Spine Surgery, Orthopedic Research Center, Imam Reza Hospital, Mashhad University of Medical Sciences, Mashhad, Iran

<sup>2</sup> Department of Orthopedics and Spine Surgery, Orthopedic Research Center, Ghaem Hospital, Mashhad University of Medical Sciences, Mashhad, Iran

Received: 07 Dec. 2016; Accepted: 16 Apr. 2017

**Abstract-** Myositis ossificans progressiva is a rare inherited disease characterized by progressive ectopic ossifications associated with thumb and big toe anomalies. Ossification usually progresses from central to the peripheral, proximal to distal, cranial to caudal, and from dorsal to ventral directions and leading to activity limitation, significant eating disability, recurrent pulmonary infection, and atelectasis. In this report, we present a 7-year-old boy with a total spine stiffness (wooden spine) seriously limited his activity of daily living.

© 2018 Tehran University of Medical Sciences. All rights reserved.

*Acta Med Iran* 2018;56(2):137-139.

**Keywords:** Myositis ossificans; Heterotopic ossification; Paraspinal muscles

## Introduction

Myositis ossificans progressiva (MOP) is a rare autosomal dominant inherited disease that usually begins in the first decade of life (1). The disease inevitably and insidiously progresses to complete stiffness and immobilization of the joints nearby. Heterotopic ossification occurs in striated muscles and surrounding soft tissues including tendons, faciae, ligaments and integumentary system (2). The disease usually affects neck, shoulder, knee and paravertebral regions while spares diaphragm, tongue, and other smooth muscles of the body (3). The most common associated anomaly seen with the disease is first toe anomaly (4). We present a case of MOP in a 7-year-old boy suffering from complete immobilization of spine from occiput to pelvis due to paravertebral muscles involvement and painful exostosis.

## Case Report

A 7-year-old otherwise healthy boy complaining of progressive stiffness and immobilization of the spine was referred to the orthopedic clinic. The disease started around a year, and before that time, the patient had no complaints at all. There was no previous history of

trauma or underlying musculoskeletal disease. The patient was from a consanguineous marriage, but no other member of the family was similarly affected.

On physical examination, the patient's spine did not have any active or passive motion. The spine was completely stiff with numerous palpable, hard masses inside the paravertebral regions of cervical, thoracic and lumbar spine. The masses were slightly tender and painful but fixed in the areas. Bilateral short thumbs and hallux valgus were also present (Figure 1). Routine blood and urine investigations (blood cell count, erythrocyte sedimentation rate, and blood chemistry values including serum calcium, phosphate, C-reactive protein, and alkaline phosphatase) were completely within the normal limits.

The radiologic investigation revealed a massive ossification throughout the spine from occiput to sacrum at the paravertebral areas (Figure 2). Mature bone completely immobilized the spine with no region left to move. We treated the patient with observation and physiotherapy as we did not aware of any better treatment modality at this time. The patient was followed-up for 6 months and during this time, the patient complains, ossification mass, pain and limitation of movement had been increased gradually. More ossification has been developed in proximal parts of the

**Corresponding Author:** F. Omid-Kashani

Department of Orthopedics and Spine Surgery, Orthopedic Research Center, Imam Reza Hospital, Mashhad University of Medical Sciences, Mashhad, Iran

Tel: +98 915 514 9248, Fax+98 51 3859 5023, E-mail address: omidif@mums.ac.ir

## Myositis ossificans progressiva

upper, and lower limbs look like exostosis.



Figure 1A and 1B. Clinical presentation of the patient



Figure 2. Radiologic presentation of the patient. Massive ossification in the lumbosacral, thoracic, bilateral scapular and cervical regions is noted

## Discussion

Myositis ossificans progressiva (also named as fibrodysplasia ossificans progressiva or Munchmeyer's disease) is a rare autosomal dominant disorder with an incidence of less than one in ten million populations that affects all races and both sexes (4). The affected gene is located on chromosome 2, and the disease frequently comes from the spontaneous new mutation. This metabolic disease is one of the three types of myositis ossificans that is characterized by the extensive metamorphosis of muscle, fascia, tendon, joint capsule, and ligament to the bone (1). Acute exacerbations of the

disease may occur at the sites of trauma, but the natural history of the disease usually begins and moves from central to peripheral, cranial to caudal and posterior to anterior manner. The disease typically spares the smooth, diaphragmatic, tongue, and cardiac muscles (3). Two main characteristics of the disease are anomalies of the thumbs and big toes, and progressive ectopic ossifications that usually begin around the age of five. Although sternocleidomastoid is frequently the sites of involvement, the disease ultimately progresses to complete bridging between any mobile parts of the body leading to wheelchair or bed confinement at the age of twenties. Eating disability, recurrent pulmonary infections, and atelectasis due to the stiffness of

temporomandibular joint and stiff chest are among the worse complications of the disease (3).

The diagnosis is usually based on the clinical features and radiological characteristics of the bony lesions. Routine laboratory tests are usually not conclusive although serum alkaline phosphatase activity, erythrocyte sedimentation rate, and C-reactive protein may be increased in flare-up phases (5). Most authors recommend that in cases with clear diagnosis, biopsy should be avoided to prevent more progression of the disease (2). MOP should be differentiated from tumoral idiopathic calcinosis, juvenile fibromatosis, lymphedema, dermatomyositis, idiopathic calcinosis, calcium metabolism diseases, and osteosarcoma (6).

Nowadays, there is no definite treatment for the disease. Corticosteroids, non-steroidal anti-inflammatory drugs, and bisphosphonates have been proposed without any effect on the disease progression (7). Any type of trauma including biopsy or surgical excision of the lesions can exacerbate the course of the disease and therefore prohibited (4). Early detection of the disease is at least helpful in protecting the child against upcoming traumas.

### Acknowledgement

We would like to show our gratitude to the Orthopedic Research Center of Mashhad University of Medical Sciences for sharing their pearls of wisdom

with us during the course of this research.

### References

1. Abdallah A, Gokcedag A, Ofluoglu AE, Emel E. Non-traumatic myositis ossificans in the lumbar spine. *Am J Case Rep* 2014;15:421-5.
2. Connor JM, Evans DA. Fibrodysplasia ossificans progressiva: the clinical features and natural history of 34 patients. *J Bone Joint Surg* 1982;64B:76-83.
3. Al-Salmi I, Raniga S, Hadidi AA. Fibrodysplasia ossificans progressiva - radiological findings: a case report. *Oman Med J* 2014;29:368-70.
4. Pignolo RJ, Shore EM, Kaplan FS. Fibrodysplasia ossificans progressiva: diagnosis, management, and therapeutic horizons. *Pediatr Endocrinol Rev* 2013;10:437-48.
5. Lutwak L. Myositis ossificans progressiva. Mineral, metabolic and radioactive calcium studies of the effects of hormones. *Am J Med* 1964;37:269-93.
6. Talbi S, Aradoini N, Mezouar IE, Abourazzak FE, Harzy T. Myositis ossificans progressive: case report. *Pan Afr Med J* 2016;24:264.
7. Illingworth RS. Myositis ossificans progressiva (Munchmeyer's disease). Brief review with report of two cases treated with corticosteroids and observed for 16 years. *Arch Dis Child* 1971;46:264-8.