

Fibrodysplasia ossificans progressiva or Munchmeyer's disease: A rare case report

ABSTRACT

Fibrodysplasia ossificans progressiva (FOP) is a rare autosomal dominant disease. It is characterized by widespread soft tissue ossification and congenital stigmata of the extremities, affecting all ethnic backgrounds. The worldwide reported prevalence is approximately 1/2,000,000. Based on history and clinicoradiological findings, FOP should be diagnosed as early as possible and noninvasively. The hallmark of diagnosis of FOP is bilateral great toe anomaly present from birth. Misdiagnosis may lead to inadvertent managements, such as manipulations, biopsies, and surgery. Surgery, till now, does not seem to have any role in the management of this condition and may lead to further trauma and disease progression. FOP may be precipitated due to trauma to muscle. In masticatory muscle, it mainly affects masseter muscle and presents with symptom of trismus. Herein, we present a case of FOP which presented to us with trismus after wooden thorn injury and immobility of the left leg. This article also emphasize on diagnosis, precaution, and treatment of disease.

Keywords: Fibrodysplasia ossificans progressiva, masseter muscle ossification, microdactyly great toe, Munchmeyer's disease, wooden thorn injury

INTRODUCTION

Fibrodysplasia ossificans progressiva (FOP) is also known as myositis ossificans progressiva (MOP), stone man disease, or Munchmeyer's disease.^[1] Myositis ossificans is mainly of two types: MOP and myositis ossificans traumatica (MOT). The worldwide reported prevalence is approximately 1/2,000,000.^[2] It was first described by Patin.^[3] The term FOP is preferred to myositis ossificans because ectopic osteogenesis occurs in the connective tissue within muscles, fasciae, ligaments, tendons, and joint capsules, rather than in the muscle fibers themselves.^[1] In masticatory muscles, however, MOT is a rare condition which was first reported by Ivy and Eby in 1924 affecting the masseter muscle.^[4] Trismus is the most frequent symptom in the masticatory muscles.^[5] Involvement of other muscles such as buccinators, medial pterygoid, lateral pterygoid, and temporalis has also been reported. In the field of dentistry, MOT can occur with any internal (e.g., local anesthetic injection, during endodontic procedure, prolonged mouth opening) and external trauma (e.g., due to trauma from external object such as thorn, pencil, and pen).^[6-9]

CASE REPORT

A 5-year-old male child came to the department of oral and maxillofacial surgery with a complaint of difficulty in opening the mouth for the past 2 years. The birth history was normal. No other member of the family was similarly affected. His mouth opening was normal until he met with a trauma with wooden thorn on the left cheek region. Thorn was removed by a surgeon immediately, but the patient had progressively

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reduced mouth opening since then. Computed tomography scan showed a radiodense mass extending in front of the anterior border of the ramus of the mandible, suggestive of ossified masseter muscle [Figure 1]. Magnetic resonance imaging revealed evidence of a large elongated T1–T2 intermediate signal intensity lesion with mild surrounding edema in the substance of left masseter muscle, abutting the ramus of left mandible likely to represent heterotopic bone formation (at the expected location of previous surgery for thorn extraction) [Figure 2]. This finding along with a history of trauma led us to the diagnosis of MOT.

On thorough probing into the history, the patient's father said that the patient had a history of pain and swelling in the left thigh region and limited movement of the left leg 3 years back when he was only 2 years old. The patient had consulted an orthopedic surgeon for further management of the same. His front and lateral leg radiograph showed bony growth over femur. Subsequent three X-rays of the leg were taken at an interval of 1 month which showed progressive increase in ossification [Figure 3a-c]. However, unfortunately, he was misdiagnosed for any bony growth in his left thigh and advised for surgical removal of the same. He got operated for the left leg in 2016; however, after operation, he had total immobility of the left leg. Postoperative radiograph of the leg showed cord-like ossification of the muscles and soft tissue of the left leg giving pattern of branching tree [Figure 4]. Local examination of the face showed that the mouth opening was reduced to nil [Figure 5]. A full-body examination revealed ossifications and a scar mark of previously operated site in the left leg regions [Figure 6]. In addition, microdactyly of the great toe on both the feet was seen [Figure 7]. Informed written consent was taken from the patient's parent.

All these clinical and radiographic features led us to conclude the diagnosis as FOP. The patient's parents were made aware of nature of disease and instructed to take possible precautions. We have kept him under observation because any surgical intervention of ossified muscle might lead to further deterioration of condition as it has been experienced by patient with two previous surgeries.

DISCUSSION

Munchmeyer's disease or FOP is a disabling genetic condition that leads to the formation of a second (heterotopic) skeleton and is the most catastrophic disorder of heterotopic ossification in humans. Throughout childhood and early adult life, this disease progressively immobilizes all of the joints of the normotopic skeleton, rendering movement impossible.^[1]

There is almost always pathognomonic microdactyly of the great toe, with suppression of the proximal phalanx. The bilateral great toe is reported in 79%–100% of patients having FOP.^[2] Heterotopic ossification in FOP progresses in specific anatomic and temporal patterns. During the first decade of life, sporadic episodes of painful soft tissue swellings (flare-ups) occur which are commonly mistaken for tumors.^[10] As the same with our case, he had developed pain and swelling of the left leg as the first complaint regarding this disease.

The etiology of FOP remains obscure. These are often precipitated by trauma, intramuscular (i.m.) injections including vaccines, local anesthesia, especially truncular block near the temporomandibular joint, muscle biopsy, and careless venipuncture.^[2] In dental, MOT can occur with any internal and external trauma, e.g., local anesthetic injection, endodontic procedure, or due to trauma from external

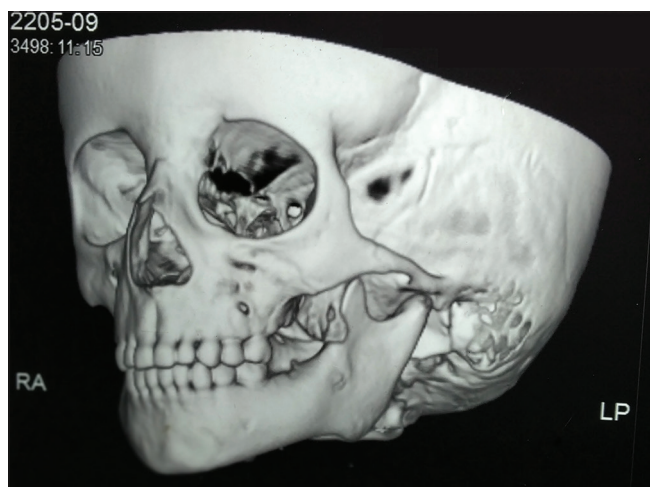


Figure 1: Three-dimensional computed tomography showing ossifying mass abutting anterior border of the mandible

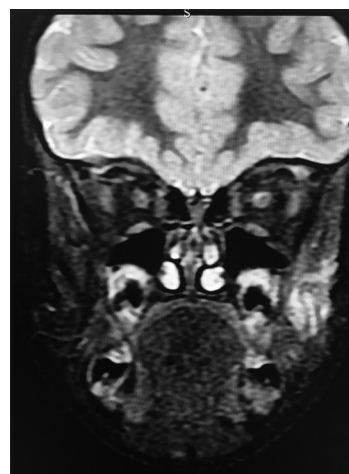


Figure 2: Magnetic resonance imaging showing mild edema surrounding left masseter and heterotopic bone formation

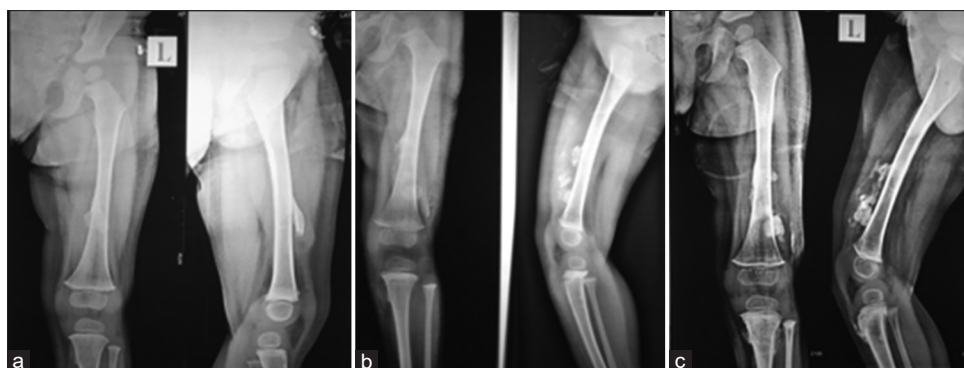


Figure 3: (a-c) Femur radiograph taken at interval dated June 17, 2016; July 21, 2016; and August 11, 2016 showing progress of ossification



Figure 4: Postoperative lower extremities X-ray showing heterotopic bone with branching tree pattern



Figure 5: Trismus and nil mouth opening

objects such as thorn, pencil, and pen.^[6-9] As wooden thorn, trauma into the left cheek and its surgical removal have led to ossification of masseter muscle in our case.

Kitterman *et al.* reported an incidence of misdiagnosis to be >90%, with 68% receiving inappropriate treatment, which leads to permanent disability in about 50% of cases.^[10] Most

patients with FOP are misdiagnosed during childhood as having cancer or aggressive fibromatosis after presentation with vascular fibroproliferative soft tissue lesions that appear in the muscles, tendons, and aponeuroses before the definitive appearance of heterotopic bone. As this happened with our case, slightly restricted movement has turned to complete immobility of the left leg after surgery and trismus after thorn removal from the left masseter muscle.

The differential diagnosis of FOP include Albright hereditary osteodystrophy, pseudomalignant heterotopic ossification, progressive osseous heteroplasia, and osteosarcoma.^[2]

Surgery, till now, does not seem to have any role in the management of this condition and may lead to further trauma and disease progression.^[3] The role of corticosteroids is uncertain and restricted to flare-ups that affect major joints, the jaw, or the submandibular area. Corticosteroids are indicated as the first-line treatment at a brief 4-day course of high-dose corticosteroids, started within the first 24 h of a flare-up, and may help reduce the intense inflammation and tissue edema seen in the early stages of the disease.^[2] Dental treatment should be carried out with utmost care peculiarly avoiding anesthesia in the mandible to prevent temporomandibular joint ankylosis.^[2] The prognosis of the disease is grave. Death may be due to respiratory infection or starvation. Starvation is caused by trismus owing to ossification in the masseter muscles.^[3]

Because of such great uncertainties regarding the treatment, we have just placed the patient under observation. Parents were advised to make child avoid sporty games, i.m. injections, arterial puncture, and physiotherapy and avoid any surgery to prevent such episodes.

CONCLUSION

Myositis ossificans is ossification and bone formation within the muscle. It is a rare and unusual pathologic entity. Early diagnosis



Figure 6: Scar mark of previous surgery and nonbending of leg also bony exostosis seen near ankle



Figure 7: Microdactyly of the great toe on both the feet

and high index of suspicion will not only prevent the iatrogenic harm but may also slow the disease progress and avoid rapid and early deterioration in patient's quality of life. Most patients with FOP are misdiagnosed early in life before the appearance of heterotopic ossification and undergo diagnostic or surgical procedures that can cause lifelong disability.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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