

Fibrodysplasia Ossificans Progressiva: TMJ Involvement and Feeding Dilemma

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ABSTRACT

Fibrodysplasia ossificans progressiva (FOP) is catastrophic uncommon hereditary connective tissue disease. The disease clinically presented as progressive ossification of ligaments, tendons as well as facial and skeletal muscles starting early in life. First presentation is painful localized soft tissue swellings. Articular dysfunction and limitation of movements is significant with involvement of the spine and proximal extremities. Limitation of mouth opening secondary to Tempromandibular joint (TMJ) involvement is late presentation of the disease. Unfortunately, surgical intervention of such condition is mentioned in literature with a very poor outcome. In this manuscript we report a case of FOP presented with limited mouth opening three months after dental extraction. Clinical findings, radiographs, and proposed treatment option are discussed.

Keywords

Fibrodysplasia ossificans progressiva (FOP), TMJ, Ankylosis.

Introduction

Fibrodysplasia ossificans progressive Munchmeyer's disease, progressive myositis ossification or Stone Mann Syndrome all are different nomenclatures of this catastrophic genetic disorder. It is a systemic connective tissue disease characterized by the formation of ectopic lamellar bone in soft tissues such as muscle, tendon, ligament, and joint capsule [1,2]. Fortunately it is a very rare type (1 every 2 million) only 800 cases documented so far worldwide. It is considered the most disabling condition in the orthopedic literature [3,4]. Mutation in the ACVR1 (bone morphogenic protein receptor) is the etiology of this disorder. At birth, the child looks normal except for congenital malformation of the great toes [5].

In the first decade, episodes of painful soft tissue swellings commonly mistaken as tumors. Some of them spontaneously regress but others will transform into bone. Usually the neck, spine, hips, shoulders, elbows and ankles are most commonly affected. At a later stage, TMJ extra articular ankylosis is evident. Usually these patients die due to pulmonary complications and chest stiffness

with subsequent respiratory failure [4,5].

Diagnosis based on history, clinical picture and radiographs (panorama, CT scans, MRI and bone scintigraphy using technetium-99m methylene radio tracer) however, definitive diagnosis is achieved by genetic testing [6]. Likely Tempromandibular joint involvement is a late presentation of the disease however it is one of morbidity factors where patients suffers from untreatable malnutrition and sever weight loss.

There is no standard therapy for this disease in the textbooks. Several medications has been postulated including NSAIDs, corticosteroids, bisphosphonates, anticoagulant therapy, rosiglitazone (Avandia), anti-diabetic drug. Radio-therapy both before and after surgery has been used. Shock wave therapy pulsed electromagnetic fields and dorsomorphin (targeting the mutant gene) all are under research [5,6].

Case Presentation

A 21-year-old female patient complained of severely restricted mouth opening three months after dental extraction under local anesthesia. The patient was diagnosed as FOP in early childhood. She had normal siblings and her parents were not consanguineous.

There were no similar conditions in her family. Orthopedics physician managed her conservatively with restriction of physical activity. Extra oral examination reveals: generalized stiffness all over her body, fixed elbows, shoulders and neck position, scoliosis (C or S shape spinal curve, frontal view, difficulty in walking which suggest hip joint affection and very short big toe. Regarding chief complain, the patient could not open her mouth more than 2-3 mm. and TMJ examination reveals minimal rotation only (Figure 1).

The patient had no history of trauma, TMJ Surgery or steroid injection. Intraoral examination could not be performed due to limited mouth opening. According to clinical findings, ankylosis was suspected and a panoramic radiograph was done, which showed bilateral TMJs degeneration and flattening of condyles. However, there was no sign of ankylosis of the TMJ (Figure 2).

For definitive diagnosis computerized tomography (CT) was done, coronal section showed flattened deformed condyles bilaterally with areas of ectopic bone formation on medial side of the joint capsule (Figure 3). Surgical intervention to release ankylosis was not advocated due to high failure rate, percutaneous endoscopic gastrostomy (PEG) tube was recommended by nutritionist to correct severe malnutrition and weight loss. However patient family refused this treatment option.



Figure 1: Clinical manifestations of FOP including A.Generalized stiffness all over the body, fixed elbows, shoulders and neck position, Scoliosis (C or S shape spinal curve), frontal view. B. Lateral view. C. Very short big toe. D. limited mouth opening.



Figure 2: Panoramic radiograph showing flattened deformed left condyle with reduced joint space. Condyle on the right side showed smaller head

with reduced joint space. poor quality of radiograph is noticed due to limited neck movement and inability to position the patient in focal trough.

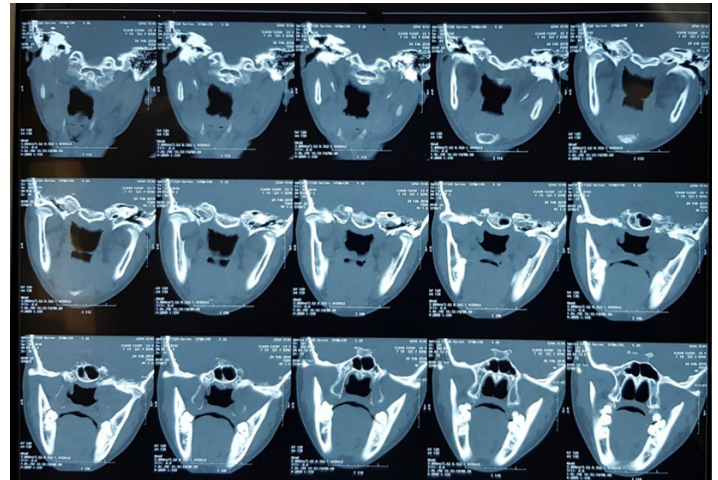


Figure 3: Coronal CT views showing bilaterally deformed condylar heads with reduced joint space and foci of ectopic bone formation in the medial aspect.

Discussion

Extra-articular ankylosis of the TMJ could be clinical presentation of myositis ossificans traumatica MO (nonhereditary heterotopic ossification) or as a part of FOP disease. Differentiation between two conditions is essential where; surgical intervention is applicable for localized form only with limited success [7]. Ankylosis of the TMJ is a common late complication in FOP cases. Our findings reinforce earlier observations that jaw involvement in FOP is correlated with age. History of trauma, dental treatment or aggressive mouth opening as mentioned in the current case raise awareness about prevention of such condition. Although orthopedic literatures could not recommend any definitive treatment of FOP, it stress on limitation of surgical interventions even diagnostic biopsy or intramuscular injection. Jiang et al hypothesized that infection and trauma exhibit an equally important role in the pathogenesis of myositis ossificans in the masticatory muscles [8].

Since ankylosis of the TMJ is a fatal un-operable complication in FOP patients, with resultant feeding difficulty and severe weight loss, prevention is better than cure. Several limitations should be considered when dealing with FOP cases including, avoid trauma even minor, muscle stretching as it may trigger the calcification. During dental treatment avoid IM (intramuscular) and IAN (inferior alveolar nerve block) or any deep injection. If general anesthesia is indicated FOP cases show difficult intubation due to neck stiffness. It's better to avoid un-necessary procedures like diagnostic biopsies and if surgery is indicated avoid the active phase of the disease. Preventive dentistry including follow-ups oral hygiene instructions and nutritional counseling is mandatory [9].

Previous reports of surgical interventions (ankylosis release) either in FOP or MO were associated with failure and case worsening. This could be due to the genetic expression of the disease, where

surgical trauma induces further extra-osseous ossifications. These clinical reports indicate the necessity of further physiologic and genetic search to clarify the etiology of FOP and provide directions to satisfactory treatment choice [10].

The case presented was suffering from severe malnutrition and weight loss where nutritional counseling was mandatory. Percutaneous endoscopic gastrostomy (PEG) tube was the recommended treatment option to correct nutritional status. However, this treatment option was not accepted by patients care givers. Percutaneous endoscopic gastrostomy (PEG) tubes are placed for a variety of conditions that interfere with a patient's oral intake. Commonly, PEG tubes are used to offer a route for enteral feeding, hydration, and medication administration in patients who are likely to have prolonged inadequate or absent oral intake however, it has reported complications as peritonitis and tube displacement [11,12].

Up to our knowledge there was no definitive treatment protocol for management of TMJ involvement in FOP cases. Muscle injury, deep injections, even minor trauma to masticatory muscles should be avoided.

Conclusion

There is deficiency in the literature about management of feeding difficulty in FOP cases. The only fact mentioned is that the outcome of the surgical intervention is not successful. Nutritionists suggest gastrostomy as only option to overcome severe malnutrition and rapid weight loss which is one of morbidity factors in FOP cases. Violation of treatment guidelines during dental treatment (deep injections and muscle stretching) could induce unwanted complication in FOP cases.

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