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Case Report

Traumatic Fibrodysplasia Ossificans: Report of Two Clinical Cases

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Abstract

Myositis ossificans is a non-neoplastic heterotopic bone formation that can affect muscles, tendons, aponeuroses, and fasciae. Its etiology is not fully elucidated yet, but it is believed that there may be a genetic predisposition from an autosomal dominant transmission, with trauma being the main triggering factor of the ossification process. Although it is most commonly found in the extremities, this condition can rarely affect the head and neck, especially affecting the chewing muscles. Given the scarcity of reports in the literature and the controversy regarding the management of these patients, this study aims to report two clinical cases attended at the Federal University of Paraná, discussing their diagnostic process and the conduct that the professional must adopt in the face of this pathology. Its presence in the maxillofacial region still needs studies and, in this context, the importance of the dentist in detecting and treating this condition stands out, which can suddenly affect the quality of life of the affected patients.

Keywords: Myositis Ossificans; Pterygoid Muscle; Computed Tomography; Trismus

Introduction

Myositis ossificans is characterized by non-neoplastic heterotopic bone formation, which can affect muscles, tendons, aponeurosis, and fascia. Its first appearance in literature was in 1648 when Patin described her patient as "the woman who turned into wood." In 1800, Bauer and Bode called it the "deformity of the petrified man". Currently, the term myositis ossificans has been gradually replaced by fibrodysplasia ossificans, once this condition is not restricted to muscles and inflammation is little evidence in affected patients [1,2].

Its progressive variant differs from the traumatic subtype, as it is an autosomal dominant disease, characterized by asymmetrical dysmorphism of the hands and feet, in addition to a progressive ossification of structures, which occurs in the craniocaudal direction, following anatomical patterns. In these patients, the temporomandibular joint will be involved in 71% of cases and most patients will be wheelchair dependent after the third decade of life [1,3,4].

Traumatic fibrodysplasia ossificans, also called circumscribed because it affects a focal point, usually have their ossification process triggered after the occurrence of acute or chronic trauma in the region. It is frequently reported in orthopedics, being in 80% of cases found in the extremities of the body. However, it is rare in the head and neck region. It has no sexual predilection and affected individuals show normal behavior and intelligence. When it occurs in the maxillofacial region, it usually affects only one site, the masseter muscle being is the most affected, probably due to its more external position on the face, making it susceptible to receiving direct trauma. Then we have the involvement of the medial and lateral pterygoid muscles and, finally, the temporal. Until 2013, only 42 cases of traumatic fibrodysplasia ossificans in masticatory muscles had been reported in the entire literature [1,2,5].

Among the factors that can trigger ossification in this group during dental practice, the following stand out: local anesthesia, especially the lower alveolar, acute or chronic trauma, surgical procedures, and facial fractures [1,6].

Although its etiology is not fully elucidated, trauma is universally accepted as a triggering agent. The first theory under study suggests that trauma generates a hemorrhage that is contained in muscle tissue, which will later ossify. Another theory involves the penetration of fragments of the periosteum, which move through the trauma towards the muscle, transporting osteogenic cells into its interior and triggering the ossification process. Several reports in the literature suggest a genetic predisposition of individuals from an autosomal dominant transmission, with varied expression [2,7,8].

Its diagnosis can be challenging for the dental surgeon, as the lesion may or may not be symptomatic. When symptomatic, its main characteristics are edema and trismus, which progress until the mouth opening is impractical. Biochemical tests, such as alkaline phosphatase and serum calcium levels, are usually within the normal range. Imaging exams vary according to the time of occurrence of the trauma. Ossification will be detectable on radiographs and computed tomography between 4 and 6 weeks after trauma, while magnetic resonance imaging can detect this process between 1 and 3 weeks with greater sensitivity. Like MRI, ultrasound can be a useful tool for early diagnosis, as it often reveals a hyperechoic zone surrounded by a well-defined hypoechoic zone before the ossifications are visible radiographically. Scintigraphy is usually used when one wants to assess the speed of progression of the ossification [2,3,5].

The biopsy is contraindicated when this pathology is suspected, as surgical manipulation can trigger a new ossification process. Its practice only gains space when the differential diagnosis involves malignant lesions, such as osteosarcoma. Otherwise, non- invasive methodologies should be adopted, such as careful clinical examination and radiographic monitoring [8,9].

Although there is no absolute consensus on the most appropriate clinical treatment, surgical excision is accepted as a way to restore the patient's quality of life. The most appropriate time for intervention is controversial, as some authors advocate early excision of the lesion (between 1 and 3 weeks) and others claim that intervention during this period increases the chance of recurrence. Several studies suggest that the interposition of material after complete excision is a good alternative to prevent bone neoformation in the region.

Abdominal fat and buccal fat pad are often used in this method. Adjuvant treatments such as bisphosphonates, non-steroidal anti-inflammatory drugs, corticosteroids, and radiotherapy have also been proposed as a way to prevent recurrences. However, there are no long-term studies that confirm the effectiveness of these resources [6].

Given the scarcity of reports in the literature and the controversy regarding the management of affected patients, this paper aims to report two clinical cases treated at the Federal University of Paraná, discussing their diagnostic process and guiding the dentist about the procedures that should be adopted in front of this diagnostic hypothesis.

Clinical case 1

A 15-year-old female patient sought care at the dentistry service in the Federal University of Paraná complaining of severe trismus. She reported that at age 12 she had undergone dental treatment, during which she received lower alveolar anesthesia on the left side. After the procedure, she felt a lot of pain in the muscles during mouth opening, which progressed to trismus. On that occasion, the professional who attended her prescribed muscle relaxants, analgesics and made a muscle relaxant plate. The limitation of mouth opening intensified. The patient was referred to the UFPR dental care service where an intervention in the joint region (she was unable to specify the procedure) was performed. The opening improved in the following 3 months after surgery, progressing with a new limitation and surgical intervention. After 30 days, the patient presented with severe trismus and no improvement in the condition.

On clinical examination, we observed total limitation of mouth opening, without pain or swelling in the face. In the tomographic exam, the three-dimensional reconstruction showed a miniplate with screws in the cervical region of the left mandibular condyle (Figure 1a), as well as the ossification of the medial and lateral pterygoid muscles (Figure 1b).

Figure 1: a: 3D reconstruction showing the presence of a mini plate with screws in the cervical region of the left mandibular condyle and b: a coronal section showing muscle ossification in the region of the medial and lateral pterygoid muscles.

Due to the proportions of the lesion, the imaging findings, and the evolution history, traumatic fibrodysplasia ossificans were suspected. On the first occasion, a biopsy was performed to rule out malignant conditions, which histologically confirmed that hypothesis.

Surgical intervention was planned to remove the ossifications through submandibular access (Figure 2a), where part of the mandibular ramus was resectioned, including the inferior alveolar nerve, and the interposition of abdominal fat was performed (Figure 2b). A remaining

stump of the mandibular condyle was left, as the temporomandibular joint remained intact (Figure 3). In other words, there were no signs of intra-articular ossification.

Figure 2: a: submandibular access for resection of ossifications and b: interposition of abdominal fat as a way to prevent recurrences.

Figure 3: Postoperative computed tomography showing the resection area, now filled with abdominal fat.

A 35 mm opening in the transoperative period was achieved and the patient was submitted to a physiotherapy regimen with daily sessions, use of muscle relaxants, and analgesics. The aperture remained stable at 25 mm for the next 3 months. Then, the intraoral infiltration of 2ml of Triamcilone (Theracort®) was started, monthly, for 10 months. During this period, mouth opening gradually decreased, until reaching 8 mm. New imaging tests were then ordered, which showed bone neoformation in the region, again limiting the mouth opening (Figure 4).

Figure 4: Panoramic radiograph showing bone neoformation in the region of the mandibular ramus, extending to the vicinity of the remaining stump.

A new surgical intervention was proposed, through the submandibular approach. After resection of the new bone mass, a titanium blade and 2.0 mm screws were used to involve the remaining

stumps, in an attempt to avoid contacts that could make recurrences possible.

Nine months after the procedure, the patient has a stable 25 mm mouth opening. The monitoring computed tomography showed bone neoformation on the titanium blade (Figure 5). So far, no new surgical intervention has been performed, but we believe that it will be necessary for the near future.

Figure 5: Side view and lingual view of the region affected by the neoformation. Its extension is observed over the titanium plate installed in the last intervention performed.

Clinical case 2

A 39-year-old male patient sought the dentistry service at the Federal University of Paraná, complaining of severe trismus and facial edema. During the anamnesis, he reported that in 1993, while practicing sports, he suffered a trauma in the posterior region of his left thigh. After not showing improvement in 3 months, affected by pain and stiffness in the affected limb, he sought an orthopedics service, where a biopsy was performed that confirmed the diagnosis of ossifying myositis. In 1994, he performed the extraction of third molars, with no reports of complications. In 2004, he began to experience pain and reduced mouth opening. On that occasion, he was prescribed corticosteroids, gabapentin, and analgesics, with no resolution of the condition. In 2011, the patient received 4 doses of Pamidorone intravenously.

At the time of care, the patient had total opening limitation (Figure 6a), with increased volume in the region of the mandibular ramus bilaterally (Figure 6b), without pain. Computed tomography showed ossification of the medial and lateral pterygoid muscles (Figure 7).

Figure 6: a: photograph of the opening measurement, showing an opening smaller than 1 mm, b: followed by changes in the proportion of the face observed during the physical examination.

Figure 7: Computed tomography showing bilateral ossification of the medial and lateral pterygoid muscles.

Surgical treatment was proposed and intubation was performed using a fiberoptic bronchoscope. Through pre-auricular, submandibular, and bilateral intraoral access, we promoted the resection of the bone masses that involved the medial and lateral pterygoid muscles. The zygomatic arches were temporarily removed to access the anterior region of the mandibular condyles, and later these were fixed with mini plates and 1.5 mm screws. Given the integrity of the temporomandibular joint, the condyles were maintained.

In the immediate postoperative period, an oral opening of 35 mm was achieved. The patient underwent a daily physical therapy regimen and infiltrations of 2 ml of Triamcinolone (Theracort®) were performed monthly. After 3 months, mouth opening was rapidly decreasing until reaching 8 mm.

Follow-up tomography showed abundant bone neoformation in the previously resected region (Figure 8).

Figure 8: Three-dimensional reconstruction showing bone neoformation in the resection area three months after the intervention.

There was a maintenance of the mouth opening at 35 mm associated with monthly infiltrations of corticosteroids bilaterally. The opening gradually decreased over time and, six months after the procedure, it is at 10 mm.

Discussion

Traumatic fibrodysplasia ossificans is a pathology that can affect muscles, tendons, aponeurosis, and fascia. It has no sexual, racial, or ethnic predilection, and may or may not manifest itself symptomatically.

Its diagnosis is challenging and, therefore, a good anamnesis is essential to identify the occurrence of acute or chronic trauma that may be related to the development of ossification. In the first case, the report of dental intervention guided the diagnostic process when correlated with symptoms and radiographic images. During the care of the second patient, the previous medical history provided evidence of the presence of the pathology, which became the main hypothesis to explain the presented condition.

One of the factors that make the diagnosis difficult is the radiographic appearance of the lesion, which changes over time and reflects the sequence of bone tissue maturation since the occurrence of the triggering factor. In addition, imaging exams that detect the development of this pathology early may involve a high cost [6].

Failure to understand its pathogenesis is also a limiting factor. In general, bone tissue has a unique capacity for growth, remodeling, and regeneration. When formed in tissues that under normal conditions would not ossify, as in fibrodysplasia ossificans, it is called heterotopic.

Several theories try to explain the atypical bone formation that occurs in these patients. Currently, the involvement of BMP-4 (Bone Morphogenic Protein) has been demonstrated in studies, since the mRNA levels of this protein are high when compared to patients without the disease. The location of the BMP-4 gene is found on chromosome 14. However, no mutations in the gene itself have been reported, suggesting that the molecular cause lies elsewhere. Furthermore, the BMP-4 signaling pathway is not fully understood [1].

Differential diagnosis includes inflammatory and infectious processes, such as thrombosis, cellulitis, osteomyelitis, and severe pericoronitis. Neoplasms such as sarcoma, osteochondroma, osteosarcoma, rhabdomyosarcoma, and metastatic diseases can also be considered, according to the proportions and behavior of the lesion [9].

Aspiration or biopsy of the central region of the lesion tends to be very cellular, which can end up leading to an incorrect diagnosis. As a rule, the biopsy is contraindicated when malignant conditions have been excluded from the scenario of hypotheses, as invasive and nonspecific procedures can trigger ossification in areas not yet affected [5].

Unlike fibrodysplasia ossificans progressive, which follows the "primum non nocere" management principle, that is, it does not primarily cause damage, the traumatic variant is usually submitted to surgical excision. This treatment modality can provide the affected patient with a quality of life, considering that severe trismus and edema can limit mouth opening. Although there is no consensus on the appropriate time to perform this intervention, it is under-

stood that early surgical excision is indicated when the injury is associated with a joint, such as the temporomandibular joint, which could generate ankylosis due to limited movement. In both cases reported, surgical treatment was able to promote a wide mouth opening in the months following the procedure, fulfilling its purpose of improving quality of life, albeit temporarily.

The use of adjuvant methods for the prevention of neoformations lacks solid scientific evidence. Bisphosphonates have been used with still uncertain benefits both in reducing pre- existing ossifications and in preventing new ossifications. Nonsteroidal anti-inflammatory drugs, radiation, local injection of corticosteroids, and physiotherapy sessions have mixed results. In the cases presented, infiltration was not able to prevent neoformation, suggesting that further studies should be carried out to identify the best treatment sequence and management of these lesions.

In addition, preventive treatment is mandatory in maintaining the oral health of these patients, to avoid performing invasive procedures that require local anesthesia or surgical interventions.

Conclusion

Attention should be paid to the differential diagnosis since it is a rare condition. If trismus and mandibular movement pain persist, this possibility should be investigated. Invasive procedures should be avoided whenever possible and, in case of severe limitation of mouth opening, procedures to improve the quality of life of the patient should be performed. Adjuvant methods to prevent relapses still need to be studied. Currently, there is a scarcity in the literature of reports involving the involvement of masticatory muscles and we hope that this work can contribute to future elucidation.

The authors disclaim any conflict of interest.

Bibliography

1. Herford AS and Boyne PJ. "Ankylosis of the jaw in a patient with fibrodysplasia ossificans progressive". *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology* 96.6 (2003): 680-684.

2. Zavattero E., *et al*. "Myositis Ossificans of the Left Medial Pterygoid Muscle: Case Report and Review of the Literature of Myositis Ossificans of Masticatory Muscles". *Craniomaxillofacial Trauma and Reconstruction* 07.01 (2013): 043-050.

3. Luchetti W., *et al*. "Severe restriction in jaw movement after routine injection of local anesthetic in patients who have fibrodysplasia ossificans progressive". *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology* 81.1 (1996): 21-25.

4. Jiang Q., *et al*. "Post-infectious myositis ossificans in medial, lateral pterygoid muscles: A case report and review of the literature". *Oncology Letters* 9.2 (2014): 920-926.

5. Godhi SS., *et al*. "Myositis Ossificans Circumscripta Involving Bilateral Masticatory Muscles". *Journal of Craniofacial Surgery* 22.6 (2011): e11-e13.

6. Trautmann F., *et al*. "Myositis ossificans traumatic of the medial pterygoid muscle: a case report". *Journal of Oral Science* 52.3 (2010): 485-489.

7. Guarda-Nardini L., *et al*. "Myositis ossificans traumatica of the temporalis muscle: a case report and diagnostic considerations". *Journal of Oral and Maxillofacial Surgery* 16 (2012): 221-225.

8. Buyse G., *et al*. "Fibrodysplasia ossificans progressiva: Still turning into wood after 300 years?" *European Journal of Pediatrics* 154.9 (1995): 694-699.

9. Cavalheiro BG., *et al*. "Multifocal myositis ossificans in masticatory muscles 30 years after gunshot wound: case report and literature review". *Brazilian Journal of Otorhinolaryngology* 85.2 (2016): 259-262.