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CASE REPORT

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Systemic scleroderma: imaging findings of diagnosis and clinical management of temporomandibular joint disorders

Esclerodermia sistêmica: diagnóstico por achados imaginológicos e manejo clínico das disfunções temporomandibulares

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ABSTRACT

Scleroderma, an autoimmune disease, directly affects the production of collagen in the connective tissue. In its systemic form, the disease causes oral manifestations such as: limited mouth opening, xerostomia, periodontal disease, thickening of the periodontal ligament and bone resorption of the mandible. This case report aims to draw attention to the difficulties encountered in providing dental care to patients with scleroderma and also to highlight the imaging findings, with emphasis on the temporomandibular joints, which are of interest to dentists about the disease. In the present case, the patient presented bilateral condylar erosion, in addition to disc displacement without reduction. Due to the systemic condition of the patient, it was decided to make an individualized occlusal splint. The limitation of mouth opening is a limiting factor for the manufacture of prostheses and plates, which is why partial prostheses are indicated and are easily removed by the patient. The decisions taken have a great impact on the health and quality of life of patients in these conditions, so there is a need for multidisciplinary involvement in order to arrive at the best treatment plan. After five years of using the stabilizing plate overnight, the patient reports greater comfort and muscle relaxation upon waking up.

KEYWORDS

Case Reports; Diagnostic imaging; Systemic scleroderma; Temporomandibular joint; Temporomandibular joint disorders.

RESUMO

Esclerodermia, uma doença autoimune, afeta diretamente a produção de colágeno do tecido conjuntivo. Na forma sistêmica, a doença causa manifestações bucais, como: limitação de abertura bucal, xerostomia, doença periodontal, espessamento do ligamento periodontal e reabsorção da mandíbula. Este relato de caso tem por objetivo chamar atenção para as dificuldades encontradas ao promover atendimento odontológico para pacientes com esclerodermia e também destacar os achados imaginológicos, com ênfase na articulação temporomandibular, que são da doença e de interessa ao cirurgião-dentista. No presente caso, a paciente apresentava erosão condilar bilateral, com deslocamento de disco sem redução. Devido à condição sistêmica da paciente, foi decidido confeccionar uma placa oclusal individualizada. A limitação de abertura bucal é um fator limitante para confecção de próteses e placas, por isso próteses parciais são indicadas, além de serem de fácil remoção pelo paciente. As decisões tomadas tem

grande impacto na saúde e qualidade de vida de pacientes nessas condições, deste modo é necessário uma equipe multidisciplinar envolvidas para chegar no melhor plano de tratamento. Após cinco anos fazendo uso da placa estabilizadora durante a noite, a paciente relata maior conforto e relaxamento muscular ao acordar.

PALAVRAS-CHAVE

Articulação temporomandibular; Diagnóstico por imagem; Escleroderma sistêmico; Relatos de Casos; Transtornos da articulação temporomandibular.

INTRODUCTION

Scleroderma is an autoimmune connective tissue disease of unknown etiology, which causes vascular damage by excessive collagen deposition. It can be classified as systemic when it affects the internal organs and skin, localized when the disease affects only the skin and muscles. It has an incidence seven times higher among women than men, usually occurring between 40 and 50 years [1]. The prevalence of systemic scleroderma (SS) varies between four and 40 cases in every 100,000 people [2]. Its appearance is rare in children and the elder [2].

The Raynaud phenomenon is one of the first signs of SS, observed in 90-98% of patients, and has characteristics of painful digital ischemia, which results in reabsorption of the terminal phalanges [3,4].

In its systemic form, in addition to compromising the muscular structure, the gastrointestinal, cardiopulmonary, and renal systems are also affected. The orofacial region in 80% of the cases presents most of the clinical manifestations [5], which starts with the rigidity of the tongue and hardening of the facial skin, presenting perioral wrinkles, which results in a classic appearance similar to a mask [3]. Some secondary manifestations, such as periodontal disease, may occur with the progression of SS [6].

Beyond the oral manifestations, characteristics such as xerostomia, thickening of the periodontal ligament and bone resorption in the mandible, limitation of mouth opening due to impairment of the temporomandibular joint, and retraction of the lips appear [4,7].

Regarding imaging exams, patients with SS have symmetrical erosions in the regions of the coronoid process, mandibular angle, and condyles [4]. Some authors report that ischemia in bone tissue associated with SS patients [8] may cause mandibular resorption.

This case report aims to raise awareness of the imaging findings of systemic scleroderma as well as demonstrate the need for a multidisciplinary team for adequate planning and management oriented to the long-term quality of the patient's life.

CASE REPORT

The Research Ethics Committee (CEP)/ Plataforma Brasil has approved this study under number: 2.002.426, the conception of the article followed the Case Reporting (CARE) guidelines for case reports [9].

In 2016, a 40-year-old female patient sought dental care claiming difficulty finding appropriate treatment in the region. During the anamnesis, the patient reported that was diagnosed with Systemic Scleroderma in 2006 at 29 years old; for dental treatment, the main complaint was dentine sensitivity; she reported difficulty performing hygiene, complained of gastric reflux, and a "tearing the skin" [*sic*] sensation during dental care.

The patient presented clinical characteristics of SS, such as a pointed nose, thin and contracted lips, perioral region of firmer consistency, and limited mouth opening (Figure 1A); at rest, the chin region had a wrinkled appearance, demonstrating that the patient must force to keep her lips closed. Thin and sensitive skin, small and slim stature, small hands with a slight curvature in all the fingers (Figure 1B) which radiographically showed the process of erosion of the distal phalanges and reduction of the remaining joint spaces (Figure 1C), on cold weather it was possible to notice the Raynaud syndrome.

In the initial panoramic X-ray, in 2016, it was possible to visualize some bone resorption of the condyle and ramus of the mandible, where bone resorption is generally more considerable

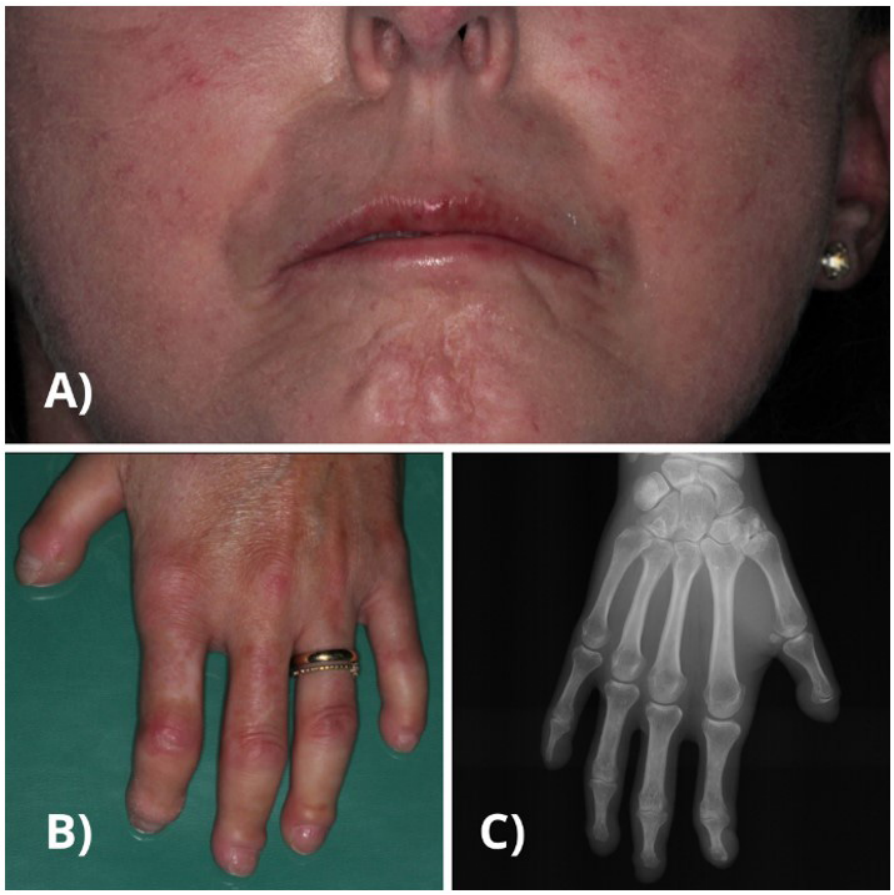

Figure 1 - (A) Frontal view of the patient, observe thin lips and contraction of the chin in resting position and narrowing of the nose; (B) Top view of the patient's left hand; (C) reabsorption in the distal phalanges.

in cases of SS. It was also possible to visualize a thickening of the periodontal ligament of some teeth (Figure 2A). Despite the thickening of the periodontal ligament, there were no clinical signs of periodontitis or occlusal trauma. It was observed gingival recession of many teeth, that justifies the main complaint of dentine sensitive of the patient, besides that, there was gingival inflammation observed during periodontal examination. The patient's diagnosis was plaque-induced gingivitis [10]. The oral hygiene instruction, scaling, and planning were conducted by sextants. The oral hygiene instruction was conducted, considering the patient's motor and mouth opening limitations, tooth extraction of the 36, and the application of 5% fluoride varnish on non-carious cervical lesions was also conducted to treat dentine hypersensitivity.

The patient remained in treatment and follow-up to control the periodontal condition. In 2018, the patient returned for care complaining of muscle discomfort in the lower third and temporomandibular joint (TMJ). A new panoramic radiograph was requested to assess the generalized oral condition when morphological changes are observed in the condyles bilaterally, hypoplastic images of the coronoid processes and an elongation of the condylar neck, and resorption of bilateral mandibular notch (Figure 2B); the patient was referred for cone beam computed tomography (CBCT) and magnetic resonance imaging (MRI) scans.

On the CBCT images (Figure 3A), on the left side, an advanced process of condylar resorption/erosion is observed throughout its extension, showing cortical irregularities, and flattening of the condylar slopes. Subchondral involvement suggests medullary necrosis, in addition, the temporal joint bone components present with cortical irregularities and bone sclerosis (Figure 3B). On the right side, there is an erosion of the condylar and mandibular fossa with discontinuity solution with consequent communication with the middle fossa of the skull. Additionally, a process of bone sclerosis is observed in the joint bone components (Figure 3C).

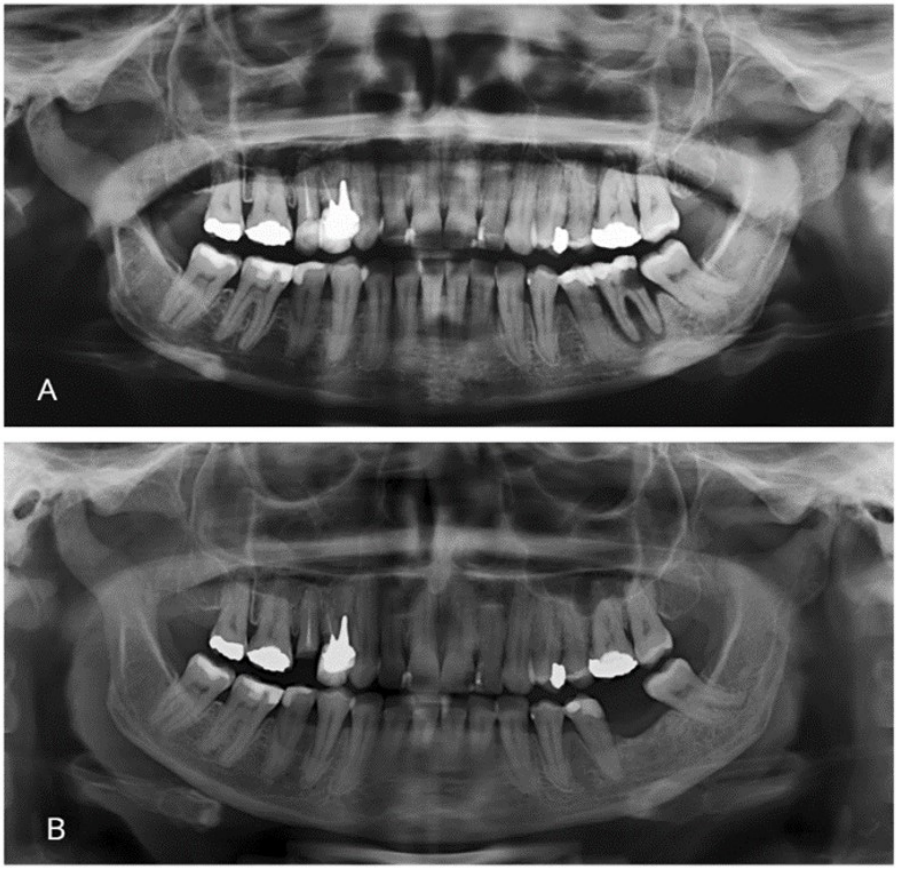

Figure 2 - A: Panoramic radiograph of the patient at the start of treatment, in 2016. B: New panoramic radiograph requested in 2018.

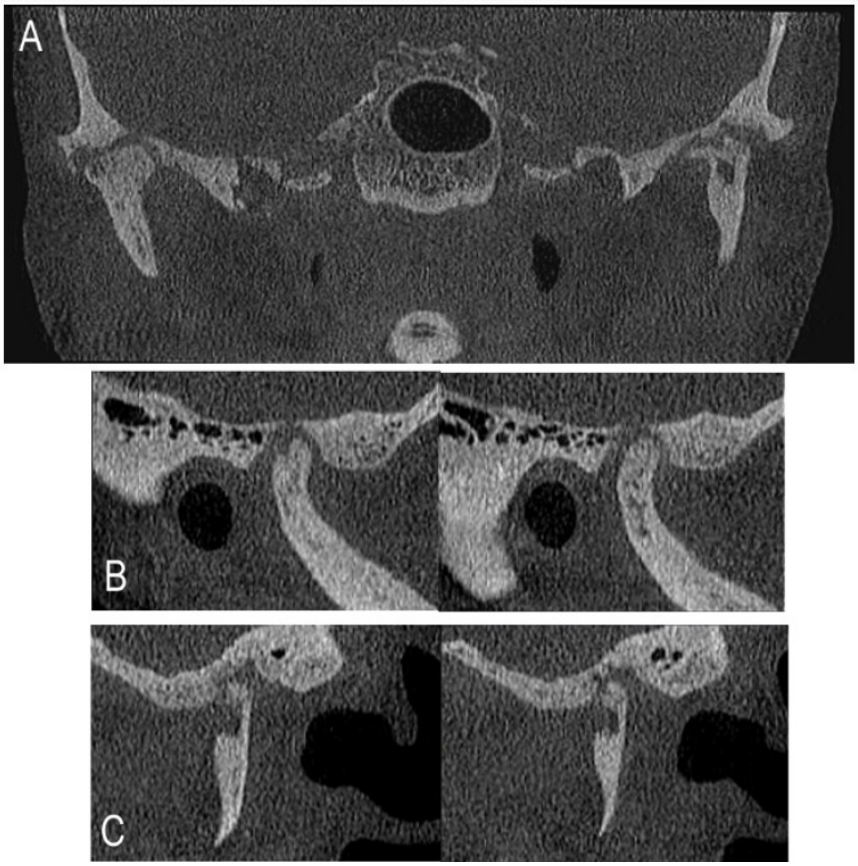

Figure 3 - A: Computed tomography showing the condition of the condyles bilaterally. B: Erosion of the right condyle with solution of discontinuity of the cortical bone, with probable dural communication. C: Computed tomography showing bone resorption in the left condyle.

At MRI, bilaterally, anterior displacement of the disc is observed with complete morphological, functional, and signal alteration (Figure 4A and 4B), as well as reduction of the joint space, fossa irregularity, and subchondral sclerosis. Also, on the left side, there is an area of hyposignal compatible with necrosis/sclerosis, suggesting a degenerative process in the condyle. Condylar morphological aspects confirm the computed tomography findings. The aspects observed in the images corroborate joint degenerative aspects.

The treatment plan for this case was based on making a stabilizing plate to relieve the internal pressure of the joints and referring the patient to an oral and maxillofacial surgeon, who, in turn, recommended the use of the plate and follow-up of the case. Because of the systemic involvement and delicate prognosis, a conservative approach to the case was preferable to performing invasive surgery.

Due to the limitation of the mouth opening, individual partial trays were made using chemically activated acrylic resin (Figure 5A) for the molding step (Figure 5B), based on the partial molds (Figure 5C) partial models were made and from them, the posterior parts of the plate were made, reduced and adapted, in the mouth; the two posterior parts were previously united with chemically activated acrylic resin, then the plate underwent a new reduction and adaptation so that it could be easily installed and removed by the patient herself (Figure 5D and 5E). After five years of using the stabilizing plate at night, the patient reports greater comfort and muscle relaxation upon waking up.

DISCUSSION

Systemic scleroderma is a rare chronic condition that affects the connective tissue, characterized by fibrosis of the skin, blood vessels, and other body systems [11]. The oral cavity is the region most involved in the pathogenesis of scleroderma, the systematic review by Benz et al. (2021) [12], demonstrated that the lips are the most affected (57.6%), followed by the oral mucosa (35.5%) and salivary glands (25.4%) [12].

Radiographically, it is possible to visualize the thickening of the periodontal ligament, more evident in the posterior teeth and present in more than one quadrant, resorption of the ramus and angle of the mandible, of the coronoid process and condyles, which occurs in at least 20% of the cases [4,11,13 -16]. Matarese et al. (2016) [17] observed that the frequency of bone alterations was significantly correlated with the mean duration of the disease [17], in the present case report, the patient showed signs of condylar degeneration twelve years after the SS diagnosis.

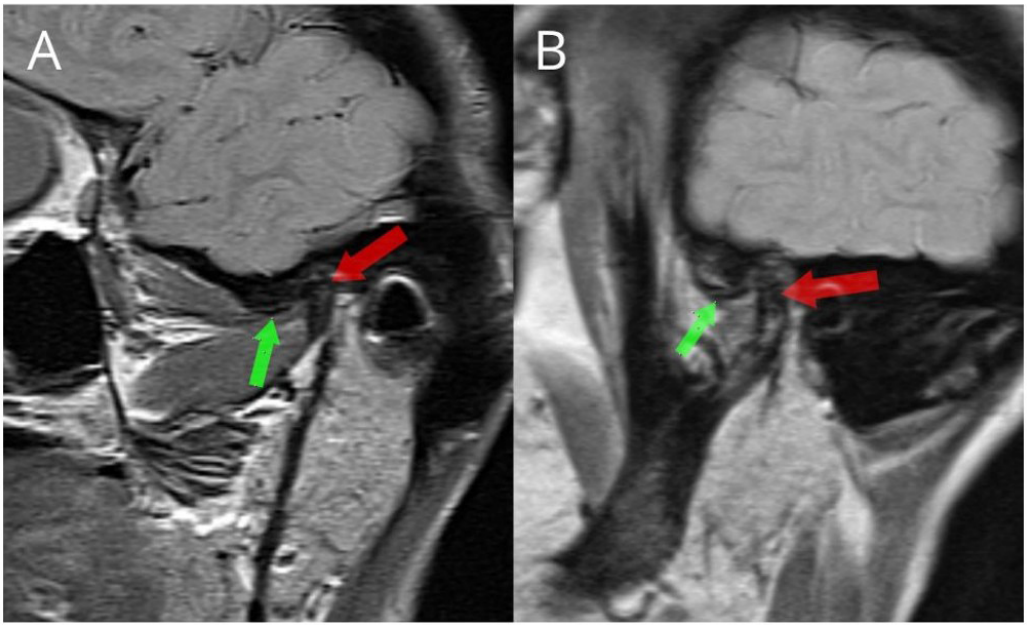

Figure 4 - T1W MRI images showing: (A) Right TMJ with joint space reduction, fossa irregularity, subchondral sclerosis, in addition to total anterior disc displacement without reduction on the right side. (B) Left TMJ with reduced joint space, fossa irregularity, subchondral sclerosis, in addition to total anterior disc reduction. Green arrows: articular disc; Red arrows: condyle.

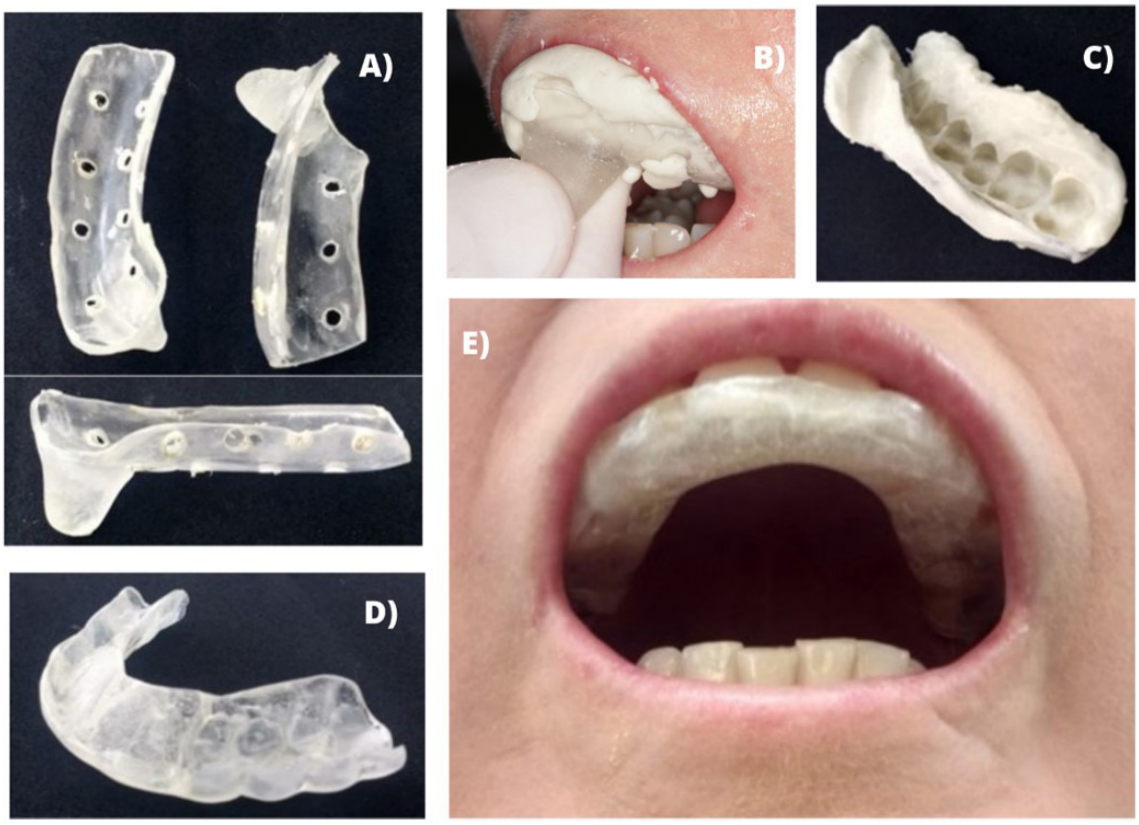

Figure 5 - A) Individual tray made of chemically activated acrylic resin for the left and right hemi-arch; B) Partial impression taken with individual acrylic resin trays; C) Partial impression of the left hemi-arch; D) Stabilizing plate complete; E) Stabilizer plate in position.

However, there is no explicit correlation between the incidence of mandibular resorption and the severity, progression, and duration of SS [18].

Regarding oral health, patients with SS have higher rates of plaque, bleeding on probing, caries, and periodontal disease. The thickening of the periodontal ligament occurs in at least two-thirds of cases of scleroderma and is considered one of the first radiographic signs that can help in the diagnosis of the disease, being associated with the increase in collagen production that occurs throughout the body [19,20]. Iordache et al. (2019) [19] verified, using CBCT, that periodontal ligament thickening is present in at least one tooth in each patient diagnosed with scleroderma, with the posterior teeth being the most affected [19]. In periapical radiographs, this thickening was 0.16 mm, twice the thickness considered normal [19].

Chebbi et al. (2016) [21], in a cohort study, observed that 30% of patients had TMJ pain, however, 90% exhibited normal radiological aspects, whereas only 10% had condylar erosion [21], therefore, TMJ pain does not necessarily indicate condylar resorption [22]. Two observational studies demonstrated that patients diagnosed with SS tend to present more signs and symptoms of temporomandibular disorders than healthy patients [17,23]. A relationship was observed between the presence of displacement of discs without reduction and bone alterations in patients with SS, possibly, this bone alteration comes from the regular absence of the disc between the condyle and temporal eminence during mandibular movement, which over time may determine alterations in the patient's sinus cavity, but this finding is not exclusive to SS [17]. Moreover, the condyle reabsorption seems to be affected by microvasculopathy and pressure ischaemia, due to skin and musculature atrophy [23].

The application of non-surgical treatments for temporomandibular disorders remains the most effective method of choice in the treatment of the patient, the treatments described in the literature include physiotherapy, application of botulinum toxin, behavioral therapy, patient re-education and awareness, and therapy with occlusal device [24]. Surgical treatment is

chosen as the last option, in the present case, considering the systemic status of the patient, non-surgical treatment was chosen, for which therapy with an occlusal splint was performed. The main idea, when using occlusal splints, is to reduce the maximum load on the TMJ, in addition to avoiding the maximum contraction of the masticatory muscles, which will promote pain reduction. In the presence of a diagnosis of SS, the limitation of mouth opening found in patients is a limiting factor for the prostheses manufacture and plates for bruxism. Therefore, it is indicated the manufacture of partial prostheses for easy removal by the patient [20].

Like Matarese et al. (2016) [17], the present case report demonstrates the need to apply imaging tests of different modalities to study the involvement of the temporomandibular joints in patients diagnosed with SS. Although studies report that the present syndrome affects the TMJ, no studies were found in the literature that used different imaging modalities to describe these alterations in the TMJs. Ideally, the dental treatment of patients diagnosed with scleroderma should be conducted individually and immediately after diagnosis. As suggestions for future work, texture analyses of the temporomandibular region could provide a more descriptive our early data about TMJ involvement in SS, since it was able to determine degenerative changes in the temporomandibular region in patients with migraine, once changes were related to the masticatory muscles and internal derangements of the articular disc [25].

CONCLUSION

The present case report indicates the importance of application of different modalities of imaging to assess TMJ degeneration and achieve an accurate diagnosis, decisions taken have a significant impact on the health and quality of life of patients in these conditions, so there is a need for multidisciplinary involvement to arrive at the best treatment plan. After five years of using the stabilizing plate at night, the patient reports greater comfort and muscle relaxation upon waking up.

Author's Contributions

LCT, ACM: Conceptualization. LCT: Methodology. CCMM, NDA, WO, ABMF, SLPCL: Methodology. CCMM, NDA, WO: Writing – Original Draft Preparation. MANJ, ABMF, SLPCL, ACM: Writing – Review & Editing. ACM: Visualization. ACM: Supervision. ACM: Project Administration.

Conflict of Interest

The authors declare that they have no conflicts of interest.

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Regulatory Statement

This study was submitted and approved by the Research Ethics Committee (CEP)/Plataforma Brasil has approved this study under number: 2.002.426.

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