Osteolysis Affecting the Jaws in Systemic Sclerosis: Clinical and Osseous Changes Based on a Case Presentation

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ABSTRACT

OBJECTIVES: The aim of the current paper is to present a case of systemic sclerosis of the jaws with all the characteristics and some extensive findings.

METHODS: Systemic sclerosis is a connective tissue condition characterized by chronic inflammatory changes, presenting with a number of symptoms. The paper aims to present a case of systemic sclerosis that had some of the characteristics of the condition unilaterally. The features were prominent and well demarcated on the panoramic radiograph.

RESULTS: The panoramic radiograph of the patient showed extensive condylar head resorption almost to the level of complete flattening.

DISCUSSION: The rarity of the condition and the awareness the oral radiologist must have upon presentation of similar images is the main reason for the presentation of the case. The features the case presents are both characteristic and well demarcated. Finally, the authors try to alert the clinician, who should be mindful of the fact that sclerodermatous involvement of organ systems is so pleotropic, that it may include the oral cavity, which is not always thoroughly observed, and is often left unattended by medical practitioners.

KEYWORDS: systemic sclerosis, scleroderma, maxillofacial findings

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Introduction

Scleroderma or systemic sclerosis is a connective tissue condition characterized by chronic inflammatory changes.[1–4] The changes include excessive collagen depositions as well as glycosaminoglycan depositions in the dermis and tissues of internal organs.[1–5]

It is of unknown etiology and often presents with a number of symptoms.

The condition is commonly identified by the presence of the characteristic Raynaud's phenomenon. The thickening of the skin presents initially with edema followed by a hardening phase with inelastic appearance and dry skin. The condition progresses with thinning of the skin and dermis and complete inelastic appearance. Collagen deposition in the vessels may cause total obliteration of the vessel leading to various other clinical manifestations, such as vasculitis of primary vessels. The maxillofacial system is affected more often by the skin condition around the mouth area and rarely by the joint involvement as in the presented case.

Case Presentation

A 54-year-old female patient presented to a private dental clinic. The main complaint of the patient upon arrival was mouth dryness (xerostomia). The medical history of the patient was taken, and systemic sclerosis was noted as a known condition. Written consent was obtained from the patient to reproduce information and photographs appearing in this work.

Upon clinical examination, the skin condition was noted, and a thorough intra- and extraoral examination revealed several major dental problems. Thus, a panoramic radiograph was suggested.

The panoramic radiograph of the patient revealed a large number of dental pathological conditions. The patient had periodontitis and caries in several teeth. The periodontal space was widened, and the widening involved the entire root surface. Besides the dental findings, bony pathological changes in the patients' jaws were noted.

The left condylar head was completely flattened and eroded and also both coronoid processes were eroded. The right condylar head had multiple small erosions on both the outer and inner poles, and a small osteophyte was present on the lateral condylar surface due to the erosions. The mandibular ramus was shortened and elevated compared to the opposite side, and slight bone loss was observed on the body of the mandible in the edentulous area on the left side.

Discussion

Systemic condition is a rare clinical entity that has been described since the 18th century; however, its etiology still remains unclear.[6]

Its prevalence is from 4 to 19 new cases per million inhabitants and mostly involves females.[7] The disease is usually present between the third and fifth decade of life, though there have been isolated reports on younger patients.[8,9] Oral manifestations are often encountered in patients with systemic sclerosis[7–20] and are summarized in the following sections.

Muscle attachment. In some cases, there have been radiographic reports of bilateral, well-demarcated, and relatively symmetrical mandibular erosions at the regions of muscle attachment, such as the angles, coronoid process, digastric region, or condyles. In this case, there was significant erosion at the region of muscle attachments and, particularly, at the condylar head, the angle of the mandible, and the coronoid processes of the mandible.[8,10,11] This resorption may be progressive in the course of the disease. Auluck et al.[12] pointed out that these erosions are caused by exerting pressure on the bone via muscular atrophy at their attachment site. This atrophy of the muscles is attributed to their increased fibrosis and a decrease in their vascularity, secondary to the fibrosis of the arterial muscular walls.[9,13] Other authors have mentioned that mandibular resorption can also be caused by ischemia of the bone secondary to the related vasculitis. The pattern of bone resorption is characteristic and is associated with the sites of muscle attachment.[14] The case presented had serious bone resorption from the edentulous side that is consistent to muscle atrophy and functional reduction together with bone resorption in the area.

Periodontal status of the condition. Rout et al.[15] reported from a series of patients that the commonest finding in the condition is widening of the periodontal ligament space and bone resorption at the sites of muscle attachment. They were the first to report root resorption at a high prevalence, 4 out of 12 patients. White et al.[16] reported bone resorption at a later stage of the condition. In the present case, there was generalized widening of the periodontal ligament. However, there was no sign of root resorption, though there was bone resorption in the edentulous area of the jaw where the forces exerted increased due to lack of teeth.

Temporomandibular joint (TMJ). Though systemic sclerosis frequently affects the TMJ, the joint is neither often described in the systemic sclerosis as affected joints nor is the condition described as a major cause of TMJ disorders. The presented case had unilateral extended mandibular condylysis, as was observed from the panoramic radiography (Fig. 1). The angles of the mandible, the coronoid processes, and the posterior borders of both ascending rami were relatively normal. The more detailed image of the condylar head, however, demonstrates the condylysis and the destructive changes in the condylar head (Fig. 2).

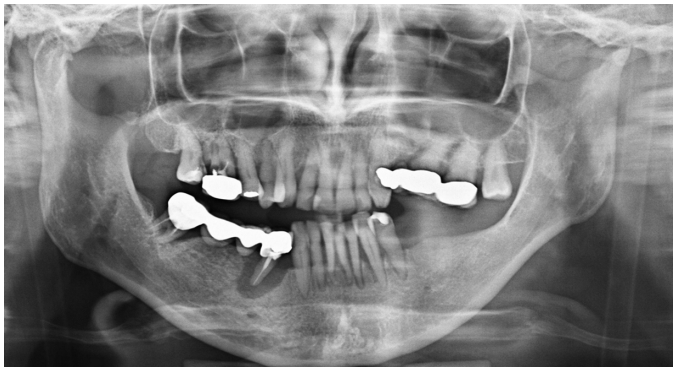

Figure 1. Panoramic radiograph of the patient.

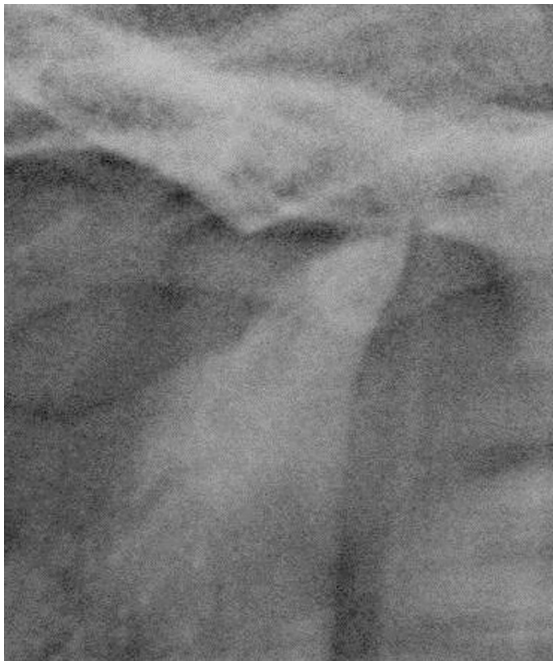

Figure 2. Magnification of the left condylar head to better demonstrate the bone condition.

In a literature review by Haers and Sailer,[17] the mandibular angle is the most commonly affected (37.6%), followed by the condyle (20.8%), the coronoid process (20.0%), and the posterior border of the ascending ramus (14.4%).

The case presented had some characteristic features of the oral manifestations of systemic sclerosis[18] and had unusually extended lesions of the condylar head.

Conclusions

Systemic sclerosis is a condition that is often unidentified in its initial stages. There have been cases with osteolysis[19] and even atrophy of the mandible.[20] Some cases may deteriorate to the extent that surgery may be needed for correction[21] The main feature that was seen in the presented case is the resorption of the mandibular condyles,[22] which may lead to all the extended pathological conditions that may accompany this condition. The radiologist should be aware of the condition

and the image characteristics of the disease and must bare it in mind in the differential diagnosis of the condition. Also the clinician who attends the patients with the condition should be mindful of the fact that sclerodermatous involvement of organ systems is so pleiotropic that it may include the oral cavity, which is not always thoroughly observed. The authors, therefore, conclude and suggest that based on the presented case and the neglection of the oral findings, early monitoring of oral changes in patients with this condition is mandatory and may improve the living conditions of the patients.

Author Contributions

Provided clinical care: EM. Analyzed the data: AD. Wrote the first draft of the manuscript: AD. Contributed to the writing of the manuscript: AD, EM. Agree with manuscript results and conclusions: AD, EM. Developed the structure and arguments for the paper: AD. Made critical revisions and approved final version: AD, EM. Both authors reviewed and approved of the final manuscript.

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