CASE REPORT

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Progressive systemic sclerosis with unilateral osteolysis of the mandible: a unique case report and review

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ABSTRACT

Objectives: This case report is aimed to discuss case findings of Progressive Systemic Sclerosis (PSS), an overview of the characteristics in the form of osteolysis on one side of the mandible, and a theoretical review.

Case Report: A 30-year old male patient came to an oral surgeon after tooth extraction. Clinical extraoral examination revealed hyperpigmentation on the right side of the face. A radiological study showed widening periodontal space on posterior teeth, and the angles of the mandibular arch, the jaw branch and the mandibular condyle neck were dissolved in the form of bone resorption.

Conclusion: Characteristics of Progressive Systemic Sclerosis (PSS) in radiographs appear in the form of expansion of the periodontal space and osteolysis of the mandibular angle, branch, and even condyle. This disease is caused by an autoimmune disease that affects the entire body, but it can manifest on one side of the body.

Keywords: *PDL widening, osteolysis lesion, progressive systemic sclerosis, scleroderma*
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INTRODUCTION

Progressive Systemic Sclerosis (PSS) is a chronic disease conventionally known as scleroderma. Scleroderma is an autoimmune disease in which connective tissue disorders and characterized by fibrosis and excessive collagen deposition in the skin, blood vessels, and internal organs such as the digestive tract, lungs, blood vessels, and kidneys. Scleroderma is more common in women, the ratio is 3-4 times that of men, and the average age of onset is about five years old. The exact etiology of systemic sclerosis is unknown. . Although genetic predisposition appears to be limited due to low genetic concordance, a familial predisposition to autoimmune disease is common.[1,2]

In the early stages of the disease, perivascular autoantibodies and lymphocytes, especially Lymphocytes T CD4+, will infiltrate the immune system and cause fibroblast activation. Lymphocytes T CD4+ cells are activated by endothelial basement membrane components such as Laminine and type IV collagen. These cells secrete endothelial cytotoxic factors called Granzyme and TNFα, which activate endothelial cells and transform them into growth factors (TGFβ), which activate fibroblasts. This condition results in the expression of TGFβ and platelet factor (PDGF). PDGF will then start the fibroblasts and then increase the collagen.[3]

The skin manifestations include the thickening of the skin, which could start with pitting edema for several months. The skin tightening replaced the pitting edema. The facial skin will appear thin and tight so that the patient will express like mask-like appearance. Oral manifestations of progressive systemic sclerosis (PSS) often include decreased ability to open the mouth, skin and mucosal pigmentation (melanoleukoderma), telangiectasia, hardening and loss of elasticity of the oral mucosa, hardening of the tongue and soft palate, xerostomia, periodontitis, difficulty chewing and speaking, widening of the periodontal ligament (PDL) and osteolysis in the form of bone resorption, especially in the mandible.[4,5]

Several characteristics are most commonly visible in radiographs, including widening of the periodontal membrane space (PDL) around the tooth, followed by thinning of the lamina dura. Because the tooth's attachment to the bone and gingiva remains normal, there is no evidence of mobility.[6,7] Furthermore, if the abnormality persists, the manifestation will continue with bone osteolysis. The mandibular angle, mandibular ramus, condyle and coronoid process, and temporal region are all common osteolysis sites which may show varying degree of bone resorption.[7,8]

The purpose of this case study is to discuss and present the findings of a case of Progressive Systemic Sclerosis (PSS), which provides an overview of osteolysis on one side of the mandible, along with a review of the theory.

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CASE REPORT

A 30-years old male patient came to a private clinic with a referral to an oral surgeon in Surabaya city. The patient's main complaint was discomfort following the extraction of tooth 26 one week ago. The extraoral examination revealed a black spot on the right cheek with tightened facial skin on the right side of the face. The intraoral mucosal examination was normal, mouth opening was standard, teeth 26 were missing, and 16 had fillings. The rest of the teeth appeared to be normal.

The results of the radiographic examination showed that there was osteolysis of the mandibular ramus that ran from the neck of the condyle thereby eliminating the anatomy of the mandibular angle. The angle area of the right mandible disappears making the right mandible a flat plane. On the left face there is no similar deformity. Based on the results of the radiographic examination on the left side of the mandible, no similar abnormalities were seen.

Based on the available data from clinical, physical and imaging examination, the provisional diagnosis of progressive systemic sclerosis (PSS) was made. The patient is then educated and referred to a general hospital for further study in an autoimmune examination to confirm the diagnosis and other treatment.

DISCUSSION

Manifestation of Progressive Systemic Sclerosis (PSS) in orofacial tissue often appears in the form of an image with specific characteristics, such as thinning and tightening of facial skin caused by subcutaneous collagen deposition. Hee's face looks expressionless, like a mask or Monalisa's face. The affected skin area will become hypopigmented/ hyperpigmented. The initial lesions begin in areas of skin induration with hair loss and sweat gland dysfunction. As the disease progresses, these lesions will turn into burning lesions with the presence of hyperpigmented sites. In addition, fibrosis occurs in the intraoral area, resulting in a small mouth.[10,11] The characteristic manifestations of this disease are often overlooked and continue into intensive care because it is not considered life-threatening. The dentist's role in detecting PSS abnormalities should be the first door for finding cases. However, due to the scarcity of issues, it is not an important thing, making dentists lack knowledge of this disease.[12,13]

From a radiological point of view, the most common picture is a widening of the periodontal ligament (PDL), which is usually found accidentally during diagnostic X-rays.[14] The incidence of widened periodontal space with intact or thinned lamina patients with PSS is about 10-37%.[6]

Figure 1. Clinical extraoral presentation of the patient shows that the skin on the right side of the face has darker pigmentation (melanoleukoderma), and the facial skin around the pigmentation appears tight[9]

Figure 2. The OPG of the patient showed osteolytic resorption at the angle of the mandible, ramus, and part of the neck of the condyle (green arrow), on the right side of the mandible, while the left mandible appeared normal. The other picture shows the widening of the periodontal space between the posterior teeth of the right mandible (red arrow)[9]

Figure 3. The periodontal space of the posterior teeth is enlarged and the lamina dura is thinned[12]

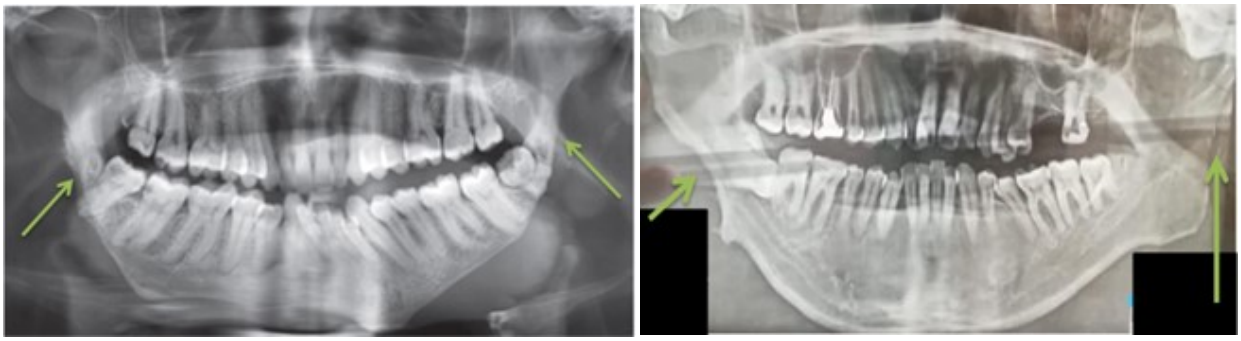

Figure 4. Osteolytic features in the form of resorption at the angle of the mandible and the mandibular ramus in two different cases[9,10]

The widening of the periodontal space (PDL) is the primary trauma of the teeth caused by the occlusal load caused by the enlarged masticatory muscles. In addition, there is an increase in collagen synthesis in the periodontal ligament. This collagen deposition requires space, which may cause the surrounding cortical bone resorption in the lamina dura so that the lamina dura thins.[14,15]

Other features commonly found on radiographs are osteolysis in the form of bone resorption and tooth-root resorption.[12,13] Osteolysis, atrophy, or resorption are common in the condyle and coronoid processes, posterior border of the ramus, and the angle of the mandible. Even in some studies, it was that PSS malignancies could also cause TMJ abnormalities and fractures.[10,11] The pathophysiology behind the skeletal changes is believed to be due to arterial muscle fibrosis, which leads to a decrease in muscle blood vessels, leading to an increase in muscle fibrosis. This fact is then followed by muscle atrophy at the site of attachment. The atrophied muscles put pressure on the bone, so the process of osteolysis begins by giving a picture such as erosion and resorption in the mandible.[10,14,15]

The mandibular ramus and the mandibular angle can be resorption because of this case. The muscle was a tension of the surface skin, resulting in muscle atrophy or bone ischemia caused by vasculitis.[5,16] According to reports, approximately 10-33% of PSS patients will experience mandibular resorption; the most common is the mandibular angle area, followed by the condyle, coronal process, and ascending branch.[17]

This resorption usually occurs on both sides of the jaw, including mandible act, ramus, then condyle. The resorption pattern indicates that the resorption occurs at the muscle insertion site.[18]

In the cases found, some changes through panoramic radiographs. The difference was in widening the periodontal membrane space, mainly in teeth 47 and 45. This widening picture conforms to the characteristics of cases of PSS patients. Another characteristic was osteolysis in the form of bone resorption. The resorption of bone includes the angle of the mandible, ramus, and neck of the condyle. This situation is also characteristic of the manifestations of PSS in the jaws and bones. Another characteristic there was hyperpigmentation on the right of the patient's cheek. These three characteristics ensure that this case is a case of Progressive Systemic Sclerosis (PSS), where there is the widening of the periodontal membrane space, osteolysis of the bone, especially at the angle of the mandible, and hyperpigmentation of the skin.[16,19]

This case was unique because the disease only occurs on one side, which is very rare. In some literature, the symptoms of the PSS in oral tissues always occur on both sides because this disease is an autoimmune disease that attacks the entire body.[17] However, in this case, it only occurs on one side, the right side. The etiology of this situation, still not explained. This situation casts doubt on the patient's final diagnosis. Unfortunately, the examination of this case could not be continued

because there were no further reports from the patient's care.

CONCLUSION

The radiographs in this case report had similar characteristics of progressive systemic sclerosis (PSS), such as widening the periodontal membrane space (PDL) and osteolysis of bone resorption, especially in the angle of the mandible ramus or condyles. Clinically, this disease can affect the appearance of the skin, like tightening and hyperpigmentation. This disease occurs due to autoimmune disorders and usually occurs throughout the body, but there are cases where the condition only manifests on one side of the body.

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FOOTNOTES

All authors have no potential conflict of interest to declare for this article. Informed consent was obtained from the patient for being included in this case report.

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