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CASE REPORT

SIGNIFICANCE OF ORAL RADIOGRAPHIC FINDINGS IN DIAGNOSIS OF PROGRESSIVE SYSTEMIC SCLEROSIS

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Abstract
Scleroderma is a chronic incapacitating multisystem disease which is an outcome of organ-based vasculopathy and fibrotic complications. Though it is rare but has the highest case-specific mortality and serves as a reminder that much of the morbidity of autoimmune and inflammatory disease occurs due to fibrosis and resultant scarring. The clinical manifestations can be divided into those that are generally present in all cases, to a greater or lesser degree, and the features only present in a minority of affected individuals. We report here a case of progressive systemic sclerosis with characteristic radiographic findings.

Keywords: Scleroderma; Lamina dura; Periodontal ligament.

INTRODUCTION

Carlo Curzio described systemic sclerosis in 1753, whereas term scleroderma was given by Gintrac in 1847.[1] The term scleroderma is derived from Greek words, “sclero” meaning hard and “derma” meaning skin, resulting to be known as “Hidebound disease”. The multiorgan damage and dysfunction result due to destruction of normal tissue architecture followed by increased deposition of extracellular matrix components. Vascular injury and activation are the earliest and possibly early events in the pathogenesis of systemic sclerosis. Female predominance is seen and majority of cases are reported in the fourth decade of life.[2] Systemic sclerosis result in considerable morbidity, depression, and hampered quality of life with increased mortality.[3] Scleroderma varies considerably in its systemic and oral manifestations from patient to patient and has an unpredictable course. The most common oral and in majority of cases initial radiographic manifestation of progressive systemic sclerosis is widening of periodontal ligament space. We attempt to discuss a case of systemic sclerosis with characteristic oro-facial and radiographic manifestations.

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CASE HISTORY:

A 36 year old female patient reported to our department with a dental complaint. Medical history revealed that she was suffering from hypertension, hypothyroidism, gastroesophageal reflux disease and interstitial lung disease and was on regular medications. She gave history of Raynauds phenomenon during winter months since five years. There was positive history of photosensitivity. General physical examination showed digital ulcers, "salt pepper" type of pigmentation and stiffening of skin. Oro-facial examination revealed "mouse facie" with anterior open bite (Fig 1). There was reduction in mouth opening secondary to changes in the skin. Intraoral examination showed reduction in salivation and gingival inflammation. The patient was further subjected to blood and radiographical investigations. Hematological investigations revealed presence of autoantibodies ANA, antidsDNA and AntiScl-70. Intraoral periapical radiography showed symmetrical generalized widening of periodontal ligament spaces in anterior and posterior teeth. There was no evidence of coronoid, condyle or mandibular angle resorption evident on panoramic radiograph (Fig 2 and 3). The history, clinical examination, hematological and radiographic investigations suggested the diagnosis of progressive systemic sclerosis.

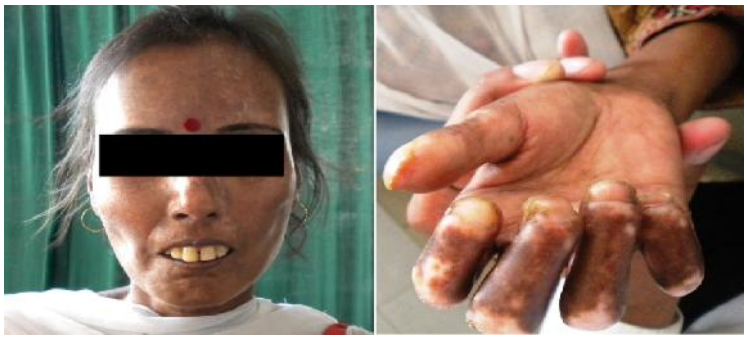

Fig 1 - Frontal extraoral view and scarring of digits of the patient

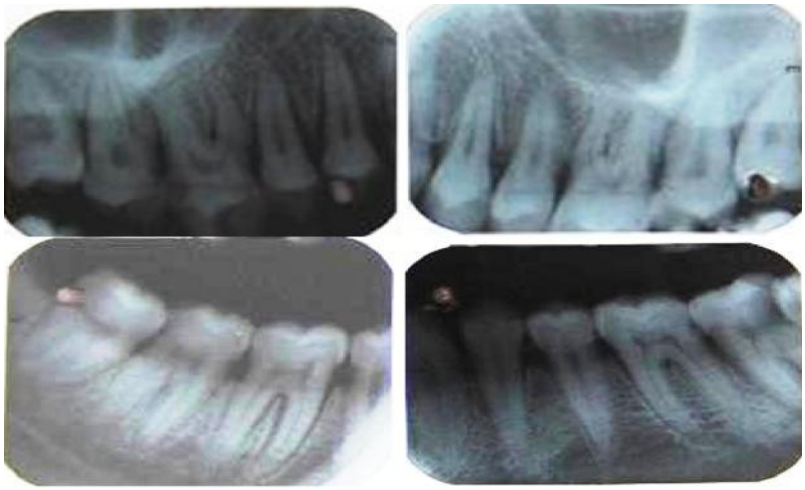

Fig 2 - Intraoral periapical radiographs showing widening of periodontal ligament space

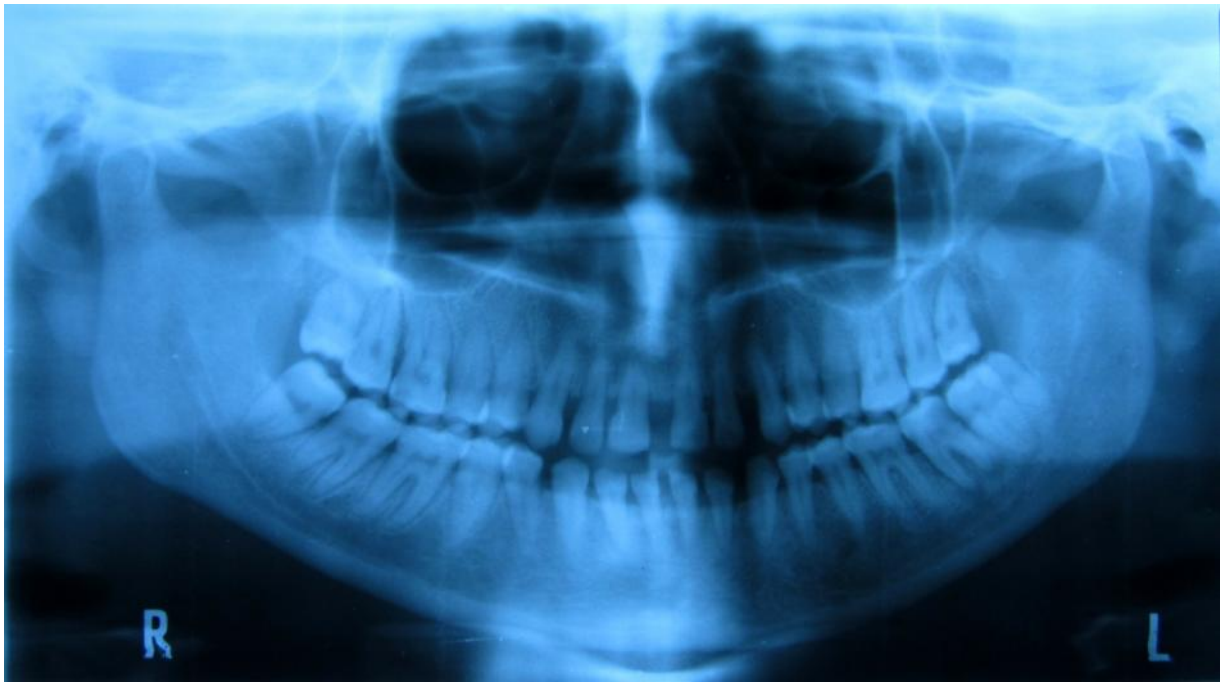

Fig 3 - Orthopantomograph showing generalized widening of periodontal ligament space

DISCUSSION:

The clinical manifestations of systemic sclerosis range from localized skin lesions affecting small areas (localized scleroderma) to systemic involvement in which multiple organ systems are affected. Localized scleroderma ropes in various types of morphea and linear scleroderma occurring in adults or children. In its classical forms, it is not associated with internal-organ disease. Diffuse systemic sclerosis designates additional widespread skin involvement proximal to the elbows, knees and/or the trunk. The initial manifestations may comprise of edematous skin, carpal tunnel syndrome, Raynaud's phenomenon, and painful joints whilst it is more likely to involve the heart, lung, kidney and viscera.

The most primitive findings in the skin may be swelling, and resultant stiffening of the joints and tendons. With time, the affected skin acquires shiny, taut, and thickened appearance with reduced elasticity and tight adherence to the underlying cutis. Concomitantly, salt-and-pepper appearance develops in skin secondary to hyperpigmentation. Similar manifestations could be appreciated in the case reported.

Around 30-50% of the patients suffering from diffuse systemic sclerosis show digital ulcers.[4] The initiation of ulcers result from failure of traumatic lesions to heal owing to fibrosis and due to compromised blood flow leading to improper tissue oxygenation along with presence of complicating factors such as infection, stretched skin over joints and epidermal thinning. The ulcers are tremendously painful and cause considerable mutilation of hand which compromise daily functions resulting in significant morbidity.[5] In our case also digital manifestations were noted thus consistent with their presence in diffuse systemic sclerosis.

The pulmonary complications include interstitial lung disease, pulmonary hypertension, pleuritis and pleural effusion, and aspiration pneumonia.[6] Patients with systemic sclerosis develop hypertension and atherosclerosis at similar or increased rates compared to those in the general population. The major cardiac complications are pericarditis, constrictive pericardium, arrhythmias, and congestive heart failure. Scleroderma renal crisis is characterized by an abrupt rise in blood pressure over days to weeks and rapidly progressive renal failure if untreated, usually within the first 5 years of the disease. It occurs virtually only in early diffuse systemic sclerosis. Gastrointestinal manifestations are common in systemic sclerosis, and the most common is esophageal dysfunction. Patients may also have dysphagia, reflux esophagitis, and the abnormal sensation of food "sticking," which necessitates drinking of fluids for relief. Rarely, telangiectasias may cause bleeding in the stomach and result in a "watermelon stomach" visible as stripes on endoscopy.[7] Pain, arthritis, tendonitis, muscle weakness, and joint contractures are common musculoskeletal manifestations. In the case presented, there was involvement of one or more organs systems.

The most commonly described oral changes are decreased opening of the mouth, microstomia, xerostomia, ulcerations and hyperpigmentation.[8] Moreover, involvement of temporo-mandibular joint apart from skin stiffness, aggravate the reduced oral opening. Complications secondary to xerostomia may also arise. Typical smooth taut mask like face result due to collagen deposition in the subcutaneous tissue of the facial skin. Oro facial manifestations such as narrowing of eyes and loss of skin folds around the mouth give a typical mask like appearance of face known as Mona Lisa face. "Mouse face" may be noted in patients with atrophy of the nasal alae. Decreased mouth opening, hyposalivation and mask like appearance were characteristically present in our case.

Most common radiographic manifestations in progressive systemic sclerosis comprise localized or generalized widening of periodontal ligament spaces around teeth with intact lamina dura which is more or less symmetrical in distribution. This characteristic radiographic manifestation has lead to diagnosis of the disease in some cases.[8] It is stipulated that increase in the connective tissue component of periodontal ligament at the cost of alveolar bone leads to widening of the periodontal ligament space evident on the radiographs. However this theory fails to justify predominance of widening in posterior teeth. Notably the study by Marmary Y et al concluded that generalized widening of periodontal ligament space was evident in all 21 cases of scleroderma but antinuclear factors were elevated in only half the patients.[9] However few researchers believed widening to be an inconsistent finding but was evident in as much as two third of the patients.[10,11] Similar widening of periodontal ligament spaces is also evident in osteosarcoma and trauma from occlusion. It is important to note here that in osteosarcoma the widening is asymmetric, more localized and lamina dura may not be intact whereas in trauma from occlusion angular bone defects are significant with accompanying mobility of teeth.[12] Other radiographic features such as bilateral, well demarcated and fairly symmetric bone erosions at the angles of mandible, coronoid process, digastric region or condyles in limited number of cases have been reported. The resorptive regions signify areas of masticatory muscles attachment which may progress in the course of the disease.[8] The radiographic evidence of generalized periodontal ligament space widening was appreciated in our case also. Conventional

and inexpensive imaging methods like panoramic and intraoral periapical radiographs are hence helpful in substantiating the diagnosis of scleroderma.

The diagnosis of systemic sclerosis is based on the identification of features that distinguish it from other autoimmune diseases, and thus a detailed history and careful physical examination are required. The American College of Rheumatology has proposed criteria to assist in identifying those affected with the condition.[13] Major criteria include scleroderma proximal to the metacarpophalangeal joints. Minor criteria are sclerodactyly, digital pitting scars, and bibasilar pulmonary fibrosis. To fulfill a diagnosis of systemic sclerosis, either 1 major or 2 minor criteria are needed.

Antinuclear antibodies have been detected in up to 90% of cases of systemic sclerosis. Anti-centromere antibodies are more likely to be associated with limited systemic sclerosis, whereas autoantibodies to topoisomerase- I (anti-Scl-70) are more likely to be associated with diffuse systemic sclerosis.[6] In the reported case also, both the antibody tests were positive.

Therapy in systemic sclerosis is directed at the organ/system involved. A 34 to 73% five year survival rate has been reported for diffuse systemic sclerosis. Male gender, older age, and involvement of the lung, kidney and heart have been associated with poorer prognosis.

CONCLUSION:

Systemic sclerosis is a rare chronic connective tissue disease of unknown etiology that can affect many organs and systems. Morbidity and mortality depend on the organ or system involved. Early recognition and diagnosis are important because effective therapies for many of the systemic manifestations are available, but timely treatment is required. As symmetric periodontal ligament space widening is classically evident in considerable number of patients suffering from progressive systemic sclerosis, general radiologists should also be made aware of its significance.

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