Progressive Systemic Sclerosis— an extensive manifestation of Scleroderma

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SUMMARY

A 32 year-old male patient, who had previously presented with all the signs and symptoms of scleroderma,was referred to a dental clinic in 2013 for removal of a lower left canine tooth that was traumatising his lower lip. His face showed tight skin with loss of wrinkling resulting in the classical "mask-like" facies. Both the sclera and intraoral soft tissues were pale, suggestive of some form of anaemia. Telangiectasia was noted in the lower lip which, together with the soft palate, was fibrosed. Mild restriction of mouth opening was noted. The patient was fully dentate but exhibited poor oral hygiene. The lower left canine was labially displaced, non-carious and firm. A panoramic radiograph showed severe resorption of the ramus, angle and body of the mandible as well as of the roots of the posterior mandibular teeth. The condyles were relatively unaffected. Multiple carious teeth were noted. The periodontal spaces appeared relatively normal. Both submandibular glands showed calcification and it appeared that there were undisplaced bilateral pathological fractures distal to the wisdom teeth. No radiological evidence of caries or periodontal disease was detected on the 33, which was extracted to relieve the patient of pain and discomfort. Healing was uneventful.

INTRODUCTION

Scleroderma, a word of Greek origin derived from the words "scleros" meaning hard and "derma" meaning skin, is a chronic inflammatory sclerotic connective tissue disorder of unknown aetiology involving the skin, muscles, lungs, heart, kidneys and gastrointestinal tract. However, a major role is thought to be played by vascular injury autoimmune factors.[1-6] Scleroderma is by no means an uncommon disease, and the purpose of this article is to present the radiographic features of a patient with progressive systemic sclerosis, a severe manifestation of scleroderma, characterised by advanced resorption of the body and ramus of the mandible together with the roots of posterior teeth. The condyles of the mandible in this case were preserved and the appearance of undisplaced bilateral fractures of the mandible distal to the wisdom teeth was noted.

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CASE REPORT

A 32 year- old male patient was referred to a dental clinic in 2013 for relief of a lower left canine tooth that was traumatising his lip. He had presented at a medical clinic during 2009 with a complaint of stiffness of the neck muscles and painful swollen joints. He was diagnosed with scleroderma having presented with thickening of the skin, multiple contractures, arthritis and sclerodactyly.

The following special investigations had been carried out: Abdominal ultrasound showed perivascular thickening and fibrosis of the liver. Loss of differentiation between cortex and medulla in the kidneys was noted. Gastric endoscopy revealed a dilated oesophagus with decreased motility. Liver enzymes were deranged. The patient had also a history of pulmonary tuberculosis complicated by a pyopneumothorax and had been on continuous home oxygen for interstitial lung disease. He was treated with mycophenolate motifel, an immunosuppressant. More recently, however, he had been diagnosed with an iron deficiency anaemia.

Clinical examination by the dental specialist showed tight skin with loss of wrinkling, resulting in the classical

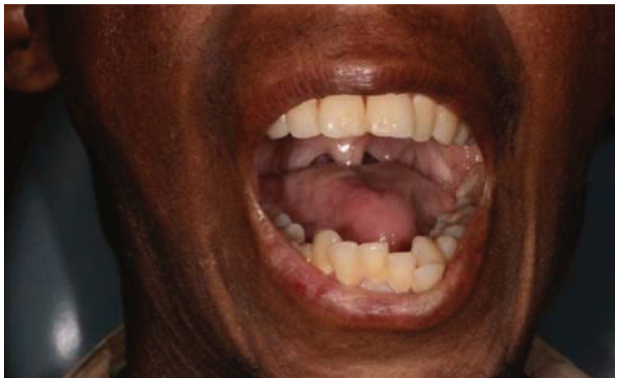

Figure 1 A: Limited mouth opening

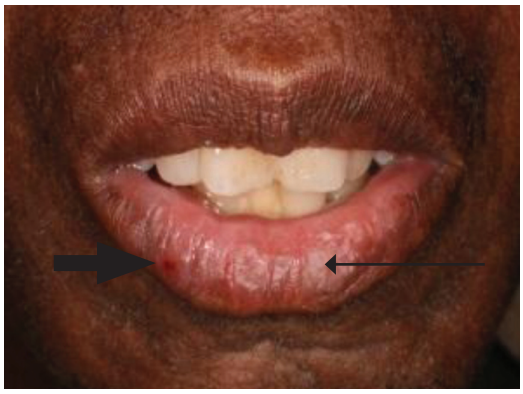

Figure 1B: Fibrosis and telangiectasia of the lower lip. Thick and thin arrows indicating telangiectasia and fibrosis respectively.

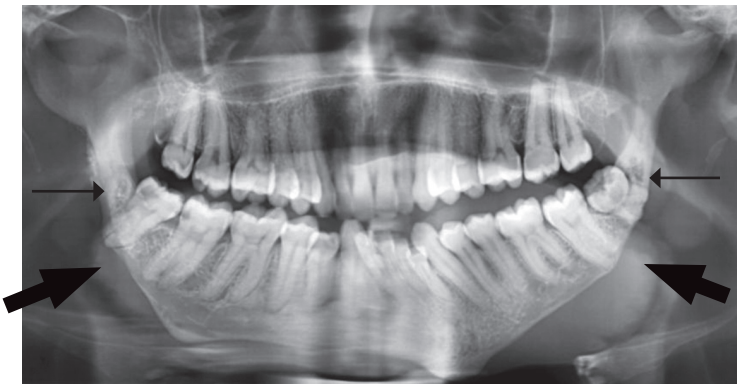

Figure 2: Panoramic radiograph showing resorption of the roots of posterior teeth, ramus, body and angle of the mandible. Thick and thin arrow indicating calcifications of the submandibular glands and bilateral pathological fractures respectively.

"mask-like" facies. Both sclera and intraoral soft tissues were pale, suggestive of some form of anaemia. The patient experienced mild restriction of mouth opening (Figure 1A). Telangiectasia was noted in the lower lip (Figure 1B) which, together with the soft palate, was fibrosed. The lip, however, displayed no neurological fallout. The patient was fully dentate but exhibited poor oral hygiene. The lower left canine was labially displaced and was reported to be the offending tooth that was traumatising the lip.

A panoramic radiograph (Figure 2) showed severe resorption of the ramus, angle and body of the mandible as well as of the roots of the mandibular posterior teeth. Neither condyle however, was affected. Multiple carious teeth were noted. The periodontal ligament spaces in this instance were within normal limits. Calcification of both submandibular glands was noted as well as the appearance of undisplaced bilateral fractures distal to the wisdom teeth. The offending 33 was non-carious and non- mobile. The tooth was extracted.

DISCUSSION

Various radiographic features have been described in patients with scleroderma.[7] These include widening of the periodontal space, resorption of the angle and ramus of the mandible, condylolysis and resorption of the roots of the teeth.[7-12]

Widening of the periodontal space with intact lamina dura is believed to occur in 10% - 37% of cases with scleroderma.[11] Increased deposition of collagen in the periodontal ligaments is thought to be the cause of the widening of the periodontal spaces. In patients where widening of periodontal spaces is the most prominent feature a differential diagnosis should include occlusal trauma as well as malignancies such as osteosarcoma

Resorption of the ramus and angles of the mandible in scleroderma is thought to be caused by pressure from taut overlying skin, atrophic muscles and ischaemia of the bone due to vasculitis associated with progressive peripheral sclerosis.[11,12] Severe resorption of the angle of the mandible often gives the radiographic impression of undisplaced bilateral pathologic fractures distal to the wisdom teeth. Resorption of roots is suggestive of long-standing pathosis.

Clinically patients present with aesthetic and facial dysfunctions which are followed by significant general and oro-facial manifestations.[1] Raynaud's phenomenon is the first presenting clinical sign followed by shortened, claw – like fingers that result from the resorption of the terminal phalanges and flexion contractions.[3,7,9,10] Abnormal collagen deposition may result in ulceration of the finger tips. The skin is termed hide-bound as a result of its developing a diffuse hard texture which is difficult to pinch and has a smooth, taut surface with loss of wrinkling. Dark pigmentation of the skin over the extremities, face and trunk may occur with contrasting areas of hypo-pigmentation.[4,9]

Oral and facial manifestations of scleroderma include para-oral and intra-oral features. Para-oral examination often reveals reduced or restricted mouth opening, "mask-like" facies and xerophthalmia. Another appearance, termed "mouse" facies is a pinched appearance of the nose that results from the nasal alae becoming atrophied. Microstomia may result from circumoral fibrosis or the lips may become pursed with radiating furrows. Telangiectasia may be present and the soft tissues around the temporomandibular joint may also be affected resulting in pseudoankylosis.

Intraoral examination often reveals that there is difficulty in maintaining oral hygiene. Rigidity of the tongue with reduced mobility together with loss of elasticity of the oral mucosa and soft palate may result in dysphonia and dysphagia. Generalized mild chronic periodontal disease, loss of attached gingiva and gingival recession is often present. Fibrotic strictures along the mandibular mucobuccal fold cause stripping of the attached gingiva. Other oral signs include mucosal pigmentation, xerostomia, increased dental decay and malocclusion.[1-9]

Three types of scleroderma (also known as hide-bound disease) have been described:

Generalized or progressive or diffuse cutaneous scleroderma results in tautness of the skin and involves the proximal and distal extremities, face, trunk, lung, heart, kidney, gastrointestinal tract and osteolytic changes in the skeleton. Localized or limited cutaneous scleroderma is limited to the distal extremities and face and also shows features of the CREST syndrome (Calcinosis, Raynaud's phenomenon, Oesophageal dysmotility, Sclerodactyly and Telangiectasia).[2,3,8,9]

Acrosclerosis is a combination of scleroderma of the extremities and Raynaud's phenomenon.[3]

The incidence of scleroderma increases with age in all ethnic groups and is three times more prevalent in females

than in males, with a peak incidence between the ages of 30 to 50 years.[1-9] The increase in prevalence may be the result of improved methods of diagnosis and/or improvements in its management resulting in patient survival.[8]

In the case of the patient described above, the offending 33 was extracted to relieve the patient of pain and discomfort and the extraction wound healed uneventfully. The less than severe restricted mouth opening was attributed to preservation to a large extent of the condyles and to minimal involvement of the muscles of mastication. This case highlights the loss of muscle tone resulting in displaced and mal-aligned teeth.

Declaration: No conflict of interest declared

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