International Journal of Community Medicine and Public Health
Kumari MV et al. Int J Community Med Public Health. 2024 Jun;11(6):2422-2426
http://www.ijcmph.com
pISSN 2394-6032 | eISSN 2394-6040

DOI: https://dx.doi.org/10.18203/2394-6040.ijcmph20241508

Case Series

Scleroderma and dental implications: a case series

M. Veera Kumari[1]*, Sumalatha M. N.[2], G. Kartheek[3], Suma Kalyani[3]

[1]Department of Oral Medicine and Radiology, Vishnu Dental College, Bhimavaram, Andhra Pradesh, India
[2]Department of Oral Medicine and Radiology, CKS Dental College, Tirupati, Andhra Pradesh, India
[3]Department of Oral Pathology, KIMs Dental College, Amalapuram, Andhra Pradesh, India

Received: 21 April 2024
Accepted: 22 May 2024

***Correspondence:**
Dr. M. Veera Kumari,
E-mail: doctorkumari1989@gmail.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

Systemic sclerosis (SSC) is a rare, systemic autoimmune disease of unknown etiology, which is characterized by fibrosis of visceral organs, skin and blood vessels. This disorder can be localized or systemic. It is more common in women with estimated prevalence is 250 cases in a million. Oral manifestations include xerostomia, periodontitis, decayed tooth etc. Radiographically generalized loss of bone with resorption of the mandibular angle and coronoid process can be evident in patients with scleroderma. Pressure of fibrous mucocutaneous tissues is thought to be the cause of the resorption. Decreased number of wrinkles due to sclerosis and distinct facial features because of the atrophy of ala nasi are among common clinical characteristics of this condition. The aim of this case series is to present two female patients with scleroderma who presented with signs of oral manifestations along with resorption at the angle of mandible, as well as widening of the periodontal space.

Keywords: Scleroderma, SSC, Crest syndrome, Xerostomia, Telangiectasia, Madonna finger

INTRODUCTION

Scleroderma is a connective tissue disorder characterized primarily by the thickening and hardening of skin. The combining form sclero means hard in Greek and the word and dermis means skin. Hidebound disease is the synonym as its characteristic feature is Hidebound skin.

Scleroderma is a chronic autoimmune sclerosing connective tissue disorder traditionally called as collagenosis, characterized by fibrosis of visceral organs, skin, blood vessels resulting from the excessive deposition of extracellular matrix components in different tissues and organs causing microangiopathy, dysregulated immune function and tissue remodeling leading to fatal complications. The disease is most common from 3rd to 4th decade of life, between the ages of 45 and 65.1

Carlocurzio of Naples in 1752 gave the first description of this disease as a separate pathological entity. Later Gintracin 1847 coined the name "sclerodermie" for the pathological entity. In 1945 Goetz coined the term "progressive SSC."[2]

Exercises such as increasing the number of tongue blades / ice cream sticks between the posterior teeth, should be encouraged. Difficulty in holding a tooth brush secondary to sclerodactyly can be overcome by advising patients to use adaptive devices such as electric tooth brushes, Waterpik-flossers and floss forks. Temporomandibular/ myofascial pain dysfunction can be treated with the use of muscle relaxants, physiotherapy and dental appliances. Dysgeusia and dental caries secondary to xerostomia can be avoided by advising the use of artificial saliva, sugar free candies, fluoride toothpaste and medications like pilocarpine.[3] In severe cases, surgical procedures, such as commissurotomy are recommended to increase mouth opening.[2] This case series makes an attempt to correlate these findings with the systemic manifestations which enable the clinician in proper diagnosis and evolve a proper treatment plan for the individual case.

CASE SERIES

A 50 and 65 year old female patient with a confirmed diagnosis of systemic sclerosis were referred to KIMS dental college hospital, from the department of dermatology and venereology, were included in the case review.

After obtaining the informed consent, patient's relevant medical history was recorded. A thorough oral examination was performed and findings were recorded. As a part of investigations, panoramic radiograph were taken and analyzed. Further, oral hygiene index simplified index (OHIS index) were also done to assess the oral hygiene status.

The patient reported with a complaint of difficulty in chewing. Patient also gave a history of pain and discoloration of her hands, overall dryness, on exposure to cold ulceration over the digits and progressive skin tightening with both hyper and hypopigmentation since 9 years. As the disease progressed, they developed restricted mouth opening, heartburn, dyspnea and altered bowel habits.

On cutaneous examination, the skin over the face (Figure 1) and extremities appeared dry and shiny. The skin when stretched showed both hyper and hypopigmentation and is hidebound in consistency. Patients presented difficulty in dressing and holding onto objects for the reason of flexion deformities of her fingers. Digital pitted scars and receded pulp of the fingers on extremities were also noticed (Figure 2).

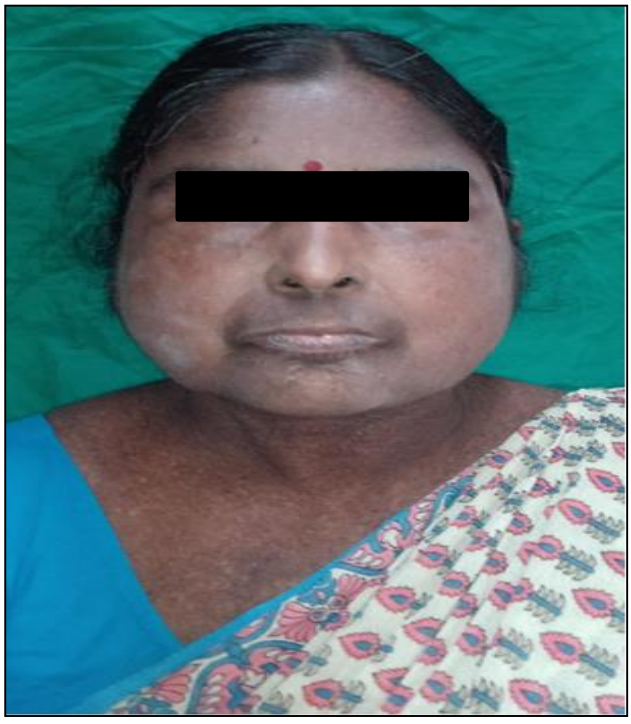

Figure 1: Facial profile.

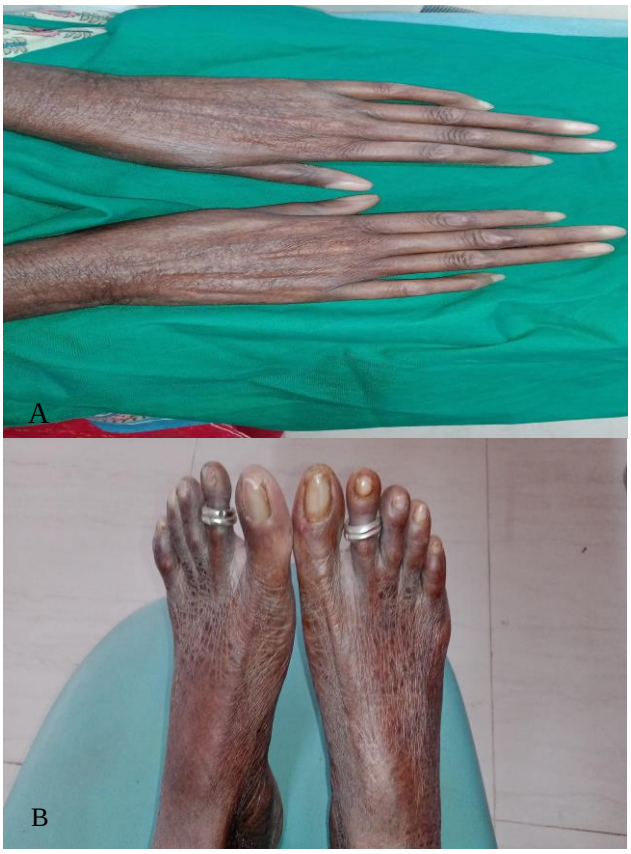

Figure 2 (A and B): Extremities.

Extra oral examination revealed loss of expression lines on the face, giving a masklike appearance, reduction of the oral aperture with the characteristic circumoral furrows (Figure 3 A and B). The lips were thinned out resembling a fish mouth with a beak shaped nose. Bilateral tenderness on tempero-mandibular joint was noticed.

Intraoral findings revealed fibrosis of the tongue with reduced mobility (Figure 4), loss of papillae, generalized pallor and blanching of the mucous membrane and diffuse fibrosis of the buccal mucosa with loss of normal elasticity (Figure 5 and 6). Interincisal distance was reduced to 3.2 cm

Periodontal findings revealed generalized gingival recession and grade I mobility, in relation to lower anterior teeth. A striking gingival finding is that 95% of the sites examined showed no gingival bleeding on probing although her oral hygiene status was poor (as demonstrated byohi-Sindex). Hypo-salivation and generalized attrition were also evident.

Patient's panoramic radiography revealed striking bilateral resorption of the angle, lower border and posterior border of ramus on right side of the mandible. Generalized interdental bone loss with symmetrical widening of the periodontal ligament space was noticed (Figure 7).

The serological reports were positive for increased serum c3 level. Ribonucleic acid (RNA) profile was positive for centromereb, Scl-70 and ro-52. A swab culture was positive for candidiasis, which reflected alteration of her oral microflora secondary to xerostomia.

Based on their history, clinical findings and further investigations the patient was provisionally diagnosed as a case of SSC. Since patients were referred to us by the dermatologist for their dental care, all patients were given basic periodontal therapy that includes; motivation, education, oral hygiene instructions, scaling and root planning. Additionally, mouth stretching exercises and facial grimacing was advised.

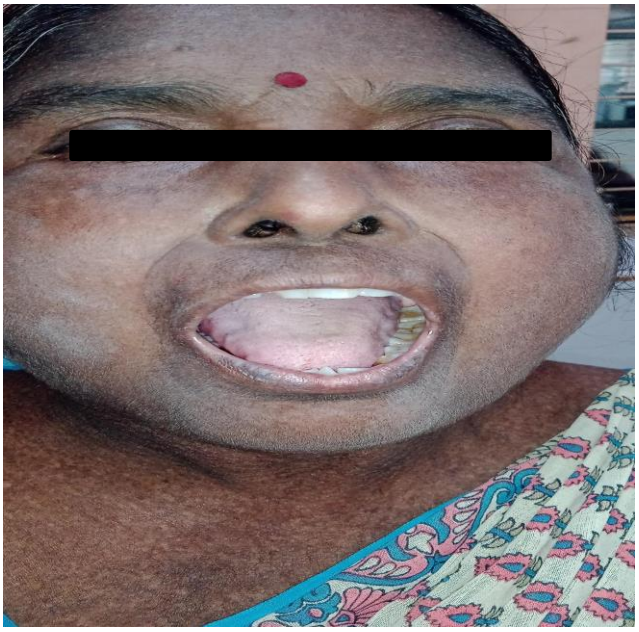

Figure 3: Decreased oral aperture.

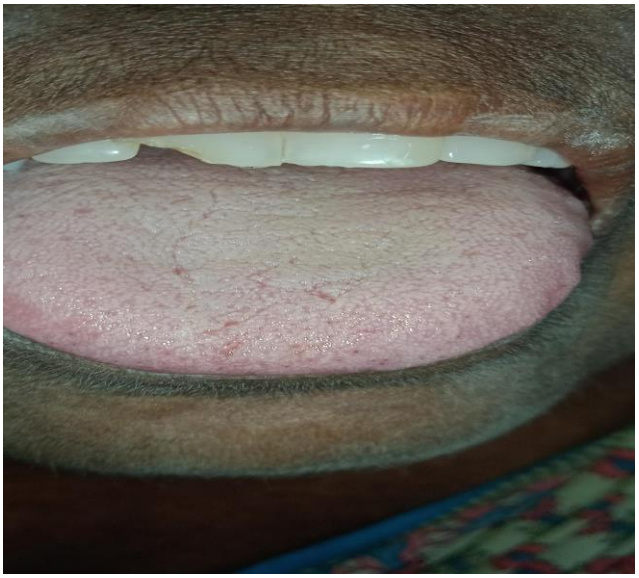

Figure 4: Restricted tongue movement.

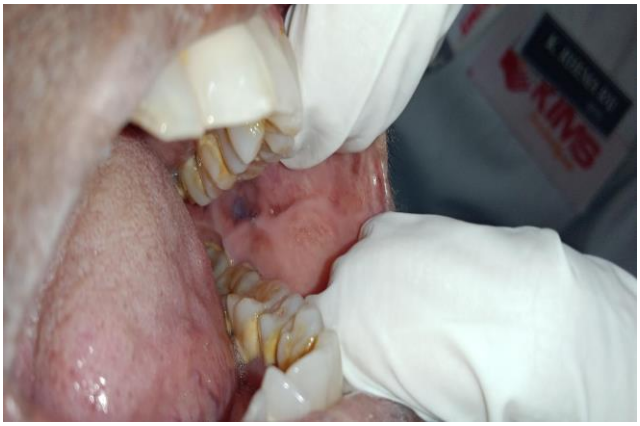

Figure 5: Left buccal mucosa.

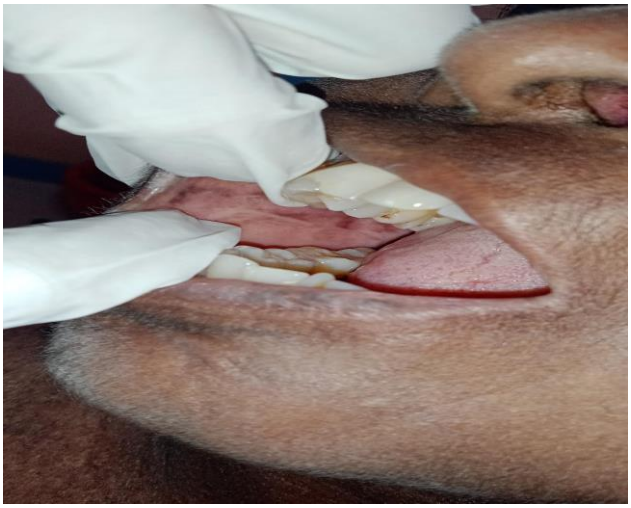

Figure 6: Right buccal mucosa.

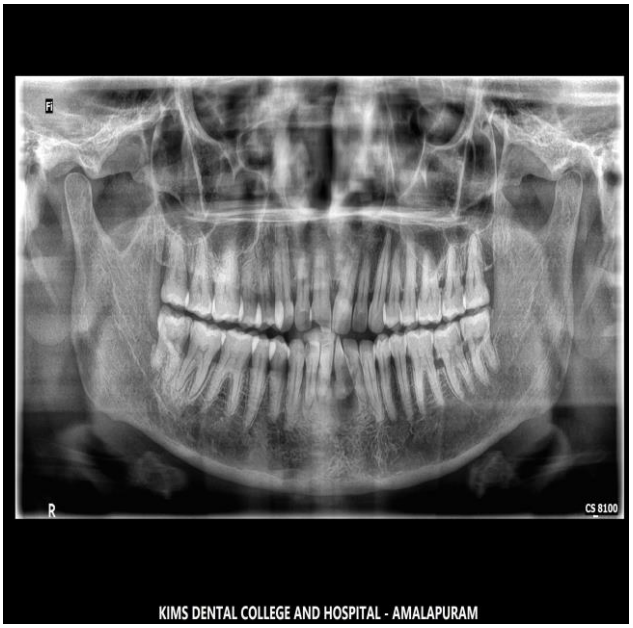

Figure 7: OPG X-ray.

DISCUSSION

SSC is a chronic autoimmune disease which poses a great challenge to clinicians with the prominent feature being progressive fibrosis resulting from excessive deposition of extracellular matrix components in different tissues and organs. Vascular damage, inflammation and presence of specific auto antibodies are also characteristic for SSC. SSC affects all the organs of the body including skin and internal organs, such as lungs, heart, kidneys, musculoskeletal system and the gastrointestinal tract causing fibrosis of all the organs. The approximated prevalence of disease is 250 per million and an annual incidence of 26/million/person/ years with a strong propensity for women (7:1) is noticed.[4] The main symptom is sclerosis of the skin which can be evaluated using the modified Rodnanskin score (MRSS) and an easily detectable marker of disease activity.[5]

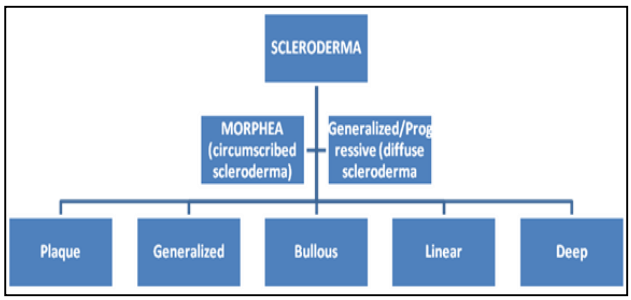

Figure 8: Types of scleroderma.

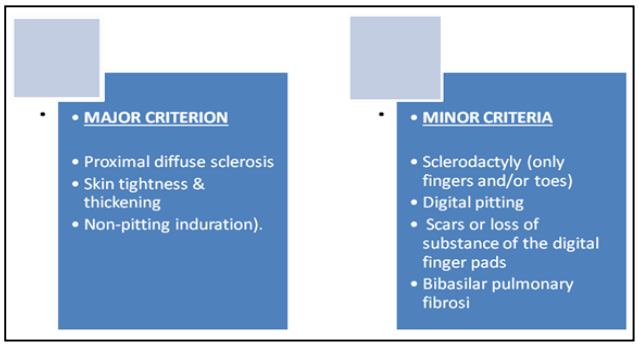

Figure 9: Criteria for the diagnosis of systemic scleroderma.

The first specific clinical sign of scleroderma is the swelling on the skin of hands and fingers "Madonna fingers".[6-8] Progressive/diffuse/generalized sclerosis is more prevalent in fourth to sixth decades of life with more female predilection.[9] The commonest oral manifestations include microstomia, xerostomia, telangiectasia, decayed teeth, severe generalized periodontitis and missing teeth. Due to increased fibrosis, tongue becomes rigid with limited movement causing speech and swallowing difficult. TMJ shows pseudo-ankylosis leading to limited mouth opening. The loss of attached gingival and gingival recession may also occur.[10] Radiographic findings include widening of periodontal space, loss of lamina dura and resorption of the Mandibular angle.

The American College of Rheumatology has recommended criteria for the diagnosis of systemic scleroderma as follows.

In the present case series, the soft tissue manifestations were similar to those reported in the earlier reports. The two cases presented here fulfils the major criteria showing symmetrical thickening of the skin proximal to the metacarpophalangeal joint (figure hands). The finding was similar to a case reported by Lester et al and few other authors.[11,12] The two individuals showed limited opening had mucogingival problems like gingival recession. The findings were in co-ordinance with cases presented by Weisman et al.[13]

The severe buccal fibrosis could be the reason for limited opening. Another striking feature in our case series is absence of gingival bleeding on probing, in-spite of poor oral hygiene. This may be attributed to vasculopathy caused by excessive accumulation of collagen in the small arteries.

The present patients demonstrated radiographic features of bone loss, erosion of mandibular ramus which were in accordance with Taveras et al who documented osteolysis of the mandibular angle and condyle in adult patients with scleroderma. These patients often suffer from severe flexion deformities of their fingers and other body joints and thus have reduced manual dexterity.[14]

The exact etiology of the osteolysis is unknown, but there are three proposed theories: Tightening of the facial skin may exert excessive pressure on the mandible and induce the bone loss; the vasculopathy associated with this disease may diminish the blood supply to the mandible resulting in bone ischemia and necrosis and atrophy of the muscles of mastication may lead to bone necrosis.

Follow-up of these patients with periodic panoramic radiographs is essential to monitor and intercept potential consequences like pathological fractures and neuropathies.[10] Crest syndrome (calcinosis cutis, Raynaud's phenomenon, esophageal dysfunction, sclerodactyly and telangiectasia), a rare condition, is thought to be a heterogeneous variant of systemic scleroderma.[15] Similarly, Parry–Romberg syndrome (hemifacial atrophy) is thought to be another variant of scleroderma.[2] In these disorders immunologically over-activated fibroblasts causes excessive deposition of normal collagen type I and III at various tissues.[16]

Treatment part includes various aspects as the etiopathogenesis of the disease is unclear. Medical management include topical intralesional or systemic glucocorticoids, vitamin E, vitamin D3, retinoid, penicillin, Griseofulvin and interferon-alpha, methotrexate, azathioprine depending upon the severity of

the disorder. Surgical intervention includes restorative plastic surgery (ferrosilicon implants, flap/pedicle grafts or bone implants).[8] Dental management includes proper maintenance of oral hygiene measures. Mouth dilators may find helpful in increasing their interincisal distance. The prognosis of the patient depends up on the course and clinical picture with characteristic organ involvement. Better collaboration between rheumatologists and the dental team is required to improve access to dental care and oral health outcomes for patients with SCC. Although early diagnosis and new therapeutic options significantly improves SC prognosis, it is still characterized by a severe course and high risk of early death. Mouth stretching exercises and facial grimacing probably are the best treatment options for this problem.

CONCULUSION

Scleroderma is a systemic disease with multi-organ involvement showing significant radiographic and clinical changes in the oral and maxilla facial structures. Oral manifestations of SSC are common and often over looked despite representing a significant cause of co- morbidity and reduced health-related quality of life in SSC. Dental professional shave the potential to identify some of the common early manifestations of this conditions like xerostomia and limited mouth opening are especially important for the right diagnosis and treatment. Multidisciplinary approach is of utmost importance for the scleroderma patients because of the complications of disease. Appropriate education on oro facial exercises and oral hygiene can be delivered by dental professionals and may significantly lessen the burden of oral manifestations in SCC. Good oral care is essential for patients with SSC helping them to keep the mouth free of dental caries and periodontal diseases. It is advised that patients with scleroderma should visit the dentist and dental hygienist at least once in three months to ensure the maintenance of good oral health care.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Jackowski J, Strietzel FP, Hunzelmann N, Parwani P, Jackowski A, Benz A. Dental implants in patient suffering from systemic sclerosis: a retrospective analysis of clinical outcomes in a case series with 24 patients. Int J Implant Dentistry. 2021;7:118.
2. Dixit S, Kalkur C, Sattur AP, Bornstein MM, Melton F. Scleroderma and dentistry: two case reports J Med Case Rep. 2016;10(1):297.
3. Bali V, Sarita D, Ashima BB, Rajiv B. A rare case of hide bound disease with dental implications. Dent Res J (Isfahan). 2013;10(4):556-61.
4. Veale et al. Oro facial manifestations of systemic sclerosis. British Dental J. 2016;221:305-10.
5. Sobolewski P, Maślińska M, Wieczorek M, Łagun Z, Malewska A, Roszkiewicz M, et al. Systemic sclerosis Multidisciplinary disease: clinical features and treatment reumatologia. 2019;57(4):221-33.
6. Dedica A, Pasalica A, Hodzicm M, Basovics S, Avdicm M, Pasice E. Scleroderma-from the aspect of dental medicine. Med Arh. 2011;65(6):368-70.
7. Altan G, Kose TE, Erdem TL, Ozcan I. Scleroderma: a case report J Istanbul Univ Fac Dent. 2015;49(2):31-4.
8. Stone J. Parry-Romberg syndrome. Pract Neurol. 2006;6:185-8.
9. Cawsonra O. Cawson's essentials of oral pathology and oral medicine. 8th ed. London: Churchill Livingstone. 2008;195.
10. Jagadishr, Mehtads, Jagadishp. Oral and periodontal manifestations associated with systemic sclerosis: A case series and review. J Indian Soc Periodontol. 2012;16:271-4.
11. Lester WB, Martinsg MJ. In: Greenberg M, Glickm, Ship JA, editors. Burket's oral medicine: Diagnosis and treatment. Immunologic diseases, 11th ed. New York: JB Lippincott publishingco. Ltd. 2008;451.
12. Anbiaee N, Tafakhori Z. Early diagnosis of progressive systemic sclerosis (scleroderma) from a panoramic view: report of three cases. Dentomaxillofac Radiol. 2011;40(7):457-62.
13. Weisman RA, Calcaterra TC. Head and neck manifestations of scleroderma. Ann Otol Rhinollaryngol. 1978;87(3pt1):332-339.
14. Taverasjm. The interpretation of radiographs in disorders of the tempero-mandibular joint. In: lschwartzed, Editor. Philadelphia: WD Saunders CO. 1959.
15. Nevillebw BW, Dammd DDd, Allencm CM, Bouquotje JE. Oral and maxilla-facial pathology. 3rd ed. Stlouis: Elsevier/Saunders. 2009;799.
16. Albilia JB, Lam DK, Blanas N, Cloakie CM, Sándor GK. Small mouths big problems? Are view of scleroderma and its oral health implications. J Can Dent Assoc. 2007;73(9):831-6.

Cite this article as: Kumari MV, Sumalatha MN, Kartheek G, Kalyani S. Scleroderma and dental implications: a case series. Int J Community Med Public Health 2024;11:2422-6.