

Rare Cases of Hide Bound Disease With Dental Implications

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Abstract: Scleroderma or Hide bound disease is a rare autoimmune disease of the connective tissue characterized by fibrosis and thickening of various tissues. Prevalence of the disease is approximately 2.7 cases per 100,000 persons per year, and orofacial involvement in localized scleroderma is reported to be 7%. This case presents with typical scleroderma features of claw deformity and mouse facies. Special dental considerations which have to be kept in mind for these patients are discussed in the article

Keywords: Scleroderma, mouse face, collagen

INTRODUCTION

Scleroderma is a chronic inflammatory disease of unknown origin and is considered autoimmune disease of the connective tissue characterized by excessive deposition of collagen and glycosaminoglycans in connective tissue of the dermis and internal organs leading to fibrosis in the skin and internal organs, resulting in thickening and hardening of the involved areas[1]. Scleroderma (Gr. *skleros*, hard, and *derma*, skin) is a generic term, used to describe both systemic as well as more localized cutaneous disorders[2].

When the systemic nature of the disease became evident, the name progressive systemic sclerosis is proposed. Though the term scleroderma is indelibly etched in the literature through common usage, the disease is currently called as “systemic sclerosis”. Since hidebound skin is the clinical hallmark of the disease, it is also called “hidebound disease”[3].

Disease course, severity, and organ involvement are variable from patient to patient, and overlap syndromes with other autoimmune conditions can occur. The pathogenesis of this disease is thought to involve three processes:

- 1) Endothelial cell damage and intimal hyperplasia
- 2) Fibroblast activation and overproduction of extracellular matrix components
- 3) Stimulation of the immune response, leading to production of autoantibodies[4].

The involvement of the skin together with the quality of its mobility, particularly in the distal portions of the extremities, is by far its most obvious symptom.

An early indicator of systemic sclerosis is Raynaud’s phenomenon which is also seen in other diseases like SLE, rheumatoid arthritis, Ehlers Danlos syndrome, etc. Thus should be differentiated from them properly, which is characterized by a painful digital ischemia, which results in local resorption of terminal phalanges. Survival of scleroderma patients is determined by the severity of visceral involvement[5].

A variant of this disease is known as the “CREST SYNDROME”, which is an acronym for calcinosis cutis, Raynauds phenomenon, Esophageal dysmotility with dysphagia, Sclerodacty and Telangiectases[6].

CASE REPORTS

Authors here present two female patients diagnosed with scleroderma having similar oral findings.

CASE I: A 50 year old medically unfit female patient diagnosed with scleroderma presented with chief complaint of burning sensation in oral cavity since 1-2 weeks on eating food. (Fig 1)

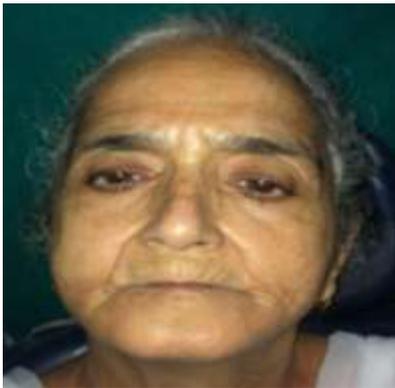


Fig 1- Mouse Facies- Thin Lips, Pinched Nose

Dental history revealed that she was edentulous since 7-8yrs, so was on soft diet since then because she was uncomfortable with the previously fabricated complete dentures.

No deleterious habits like consumption of tobacco (smoking or smokeless form) were reported.

Medical history revealed history of hypothyroidism and hypertension and was on medications for same, with additional multivitamin supplements and calcium for last 25 years.

On general physical examination, eyes were dry with mild itching and pain. Pale nails presented with vertical ridges and koilonychia. There was blue spot (telengectasia) present on plantar surface of left hand. Claw deformity could be appreciated on little fingers of hands but toes were normal (FIG 2 & 3).



Fig-2: Normal Toes



Fig-3: Pale Nails With Vertical Ridges & Koilonychia. Blue Spot (Telengectasia) Present On Plantar Surface Of Left Hand And Claw Deformity Of Little Finger On Both Hands.

Facial skin was tight and thick, wrinkles near eye and mouth were lost with very less wrinkles appreciated on forehead giving "mask like appearance" (FIG 4).

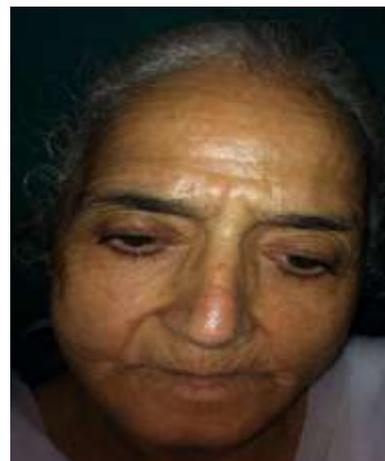


Fig-4: Mask Like Appearance- Loss Of Wrinkles, Tight And Thick Skin

The lips appeared thin and nasal alae were atrophied giving pinched appearance to the nose, classically known as "mouse facies"(FIG 1).

Intraoral examination, revealed thick frothy saliva suggestive of xerostomia. There was generalized pallor, diffuse fibrosis of oral mucosa with reduced flexibility, restricted mouth opening (FIG 5) and bald tongue (FIG 6).

After the written consent from the patient orthopantomogram was made which revealed completely resorbed edentulous ridges with soft tissue calcification bilaterally on right and left pharyngeal walls (FIG 7).



Fig-5: Reduced Mouth Opening



Fig-6: Bald Tongue

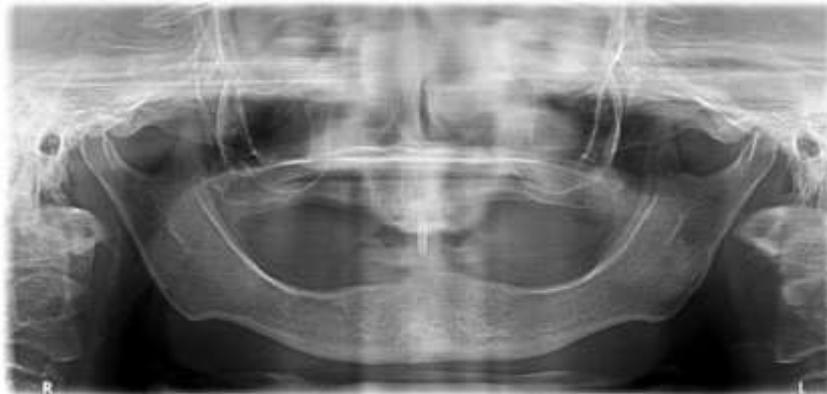


Fig-7: Soft tissue calcification bilaterally on right & left pharyngeal walls.

Patient was diagnosed having oral manifestations of scleroderma and anemia and edentulous upper and lower arches. Topical corticosteroids were prescribed to relieve the burning sensation. She was also advised for new dentures and consult physician for anemia. Her follow up was done on regular basis.

CASE II: Another 52 year old female patient diagnosed with scleroderma reported to the department of Oral Medicine and Radiology with similar extraoral and intraoral presentation (FIG 8) along with flexure contracture producing shortened 'claw like' fingers, digital pitting scars on palmar surface of hands (FIG 9,10&11).



Fig-8: Extraoral And Intraoral Presentation



Fig-9: Claw Deformity With Digital Pitting Scars



Fig-11: Digital Pitting Scars On Palmar Surface Of Hands



Fig-10: Digital Pitting Scars On Palmar Surface Of Legs

On patients informed consent, roentgenographic examination were done which revealed resorption of terminal phalanges (FIG 12) and generalized loss of lamina dura of teeth in mandibular dental arch (FIG 13).



Fig-12: Resorption Of Terminal Phalanges



Fig-13: Generalized Loss Of Lamina Dura

After a thorough biochemical investigations and physicians consent, new set of dentures were fabricated to satisfy the patients needs (FIG 14).



Fig-14: Patient With Dentures

Both the patients are under regular follow up.

DISCUSSION

Scleroderma is a rare condition, first characterized as a single condition in 1752 by Curzio of Naples. It predominantly affects woman between 30 and 50 years of age as also in the cases presented by the authors.

There are two major classifications of scleroderma: localized scleroderma (morphea & linear) and systemic sclerosis (limited & diffuse)[7].

One of the first signs of the disease is Raynaud's phenomenon (bluish discoloration of hands on exposure to cold) followed by the resorption of the terminal phalanges and flexion contractures resulting in shortened, claw like fingers as presented in these case reports. Less commonly, ulceration of the finger tips due to abnormal collagen deposition is also noted[8]. The skin develops a diffuse, hard texture which is difficult to pinch (hide bound skin) and its surface is usually smooth, taut. Skin over the extremities, faces, and trunk may become darkly pigmented and contrasting areas of hypo pigmentation may also develop. The sparing of pigment around hair follicle gives the skin a 'salt and pepper' appearance[9].

Similarly, the nasal alae become atrophied, presenting as mouse facies, same as in the cases presented. Oral manifestations include decreased opening of the mouth, microstomia and xerostomia[10].

The most common oral radiographic findings, which occurs in about two-thirds of patients, is an increase in the width of the PDL around the teeth, but the lamina dura remains normal. The condyle and the coronoid process shows resorption and painful trigeminal neuropathy caused by nerve compression. "Tail of the whale," pattern is appreciable due to blunting of angle of mandible[11].

Although the disease can be diagnosed with proper clinical examination, but radiographic and blood investigations confirms the diagnosis. As such there is no cure for the disease but proper care should be taken, to prevent further progression and should be treated symptomatically.

Some symptoms develop with relative suddenness; others take years to develop. The exact course the disease may take is unpredictable, and the prognosis will vary from individual to individual. Systemic scleroderma is a chronic, life-long disease[12].

Dental management for these patients is very important to prevent teeth loss at an early stage and other side effects of disease. These cases can be managed with following few simple steps:

1) Xerostomia

- Frequent sipping of water, minimum of 8-10 glasses of water to maintain all body functions including production & quality.
- Artificial saliva and substitutes.
- Sugar free chewing gums provide stimulation of saliva[13].
- Milk with olive oil provide lubrication to oral tissues.
- Evening primrose oil, anhydrous crystalline maltose and dehydroepiandrosterone (DHEA), moisturizing gels[3].

2) For dentulous patients

- Proper cleaning of teeth and use of dental floss.
- Electric toothbrushes in patients whose manual dexterity is impaired.
- Antiseptic mouthwashes and fluoride application[14].

3) Edentulous patients

- Flip dentures
- Sectioned dentures
- Elastic dentures
- Antifungal lozenges, drops, gels, ointments or creams to prevent candidal infection.

4) Microstomia

- Mouth exercises & facial grimacing[15].

5) Advice patient to avoid large doses of Vitamin-C because it stimulates collagen formation and may enhance deposition.

6) Stress management

- Proper sleep and rest
- Avoid stressful situations
- Eating healthy diet
- Learning methods to control anxiety & fears
- Exercising.

7) Dental examination every 3-6 months.

CARE OF SYSTEMIC INVOLVEMENT

1) Raynaud's phenomenon

- Quit smoking
- Dress warm, and keeps hands and feet warm
- Do exercises that relax body

2) Skin thickening- D-penicillin, interferon-gamma, cyclophosphamide.

3) Pruritis- moisturizers

4) Raynaud phenomenon- calcium channel blockers, aspirin, topical nitrates.

5) GI problems- antacid, H2 blockers, proton pump inhibitors, laxatives. Limit use of alcohol, spicy & acidic food.

6) Pulmonary fibrosing alveolitis- cyclophosphamide

7) Pulmonary hypertension- supplemental oxygen, Bosentan

8) Renal crisis- ACE inhibitors

9) Myositis- steroids, methotrexate, azathioprine.

10) Arthralgias- acetaminophen, NSAIDs.

11) Regular visit to physician for systemic check up[16].

LIVING WITH SCLERODERMA

Living with scleroderma is quite challenging. Everyday activities can sometimes be difficult due to physical limitations and pain. Patient must deal with psychological setbacks that come from living with the disease that is chronic, uncommon and currently incurable. Because scleroderma can cause significant changes in appearance, a patient's self-esteem and self-image are almost always affected. The support of family and friends is vital in helping to maintain a good quality of life.

CONCLUSION

Scleroderma has the potential to have a significant adverse effect upon the oral cavity. It is important that all persons with scleroderma maintain an adequate standard of oral hygiene to lessen the risk of complications of disease - particularly early tooth loss. Individuals with scleroderma are strongly advised to obtain appropriate specialist advice via their doctors or medical specialists.

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