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Scleroderma-The Review

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ABSTRACT:

The cause of the chronic connective tissue condition scleroderma is unknown to man. Improper extracellular matrix deposition in connective tissues and cells, which results in disturbances and tissue hypoxia, distinguishes it.

Skin or mucosal atrophy, subcutaneous cells or tissues, muscle atrophy, and internal organ atrophy are some of the alterations that occur.

Both morphea and diffuse alterations fall into this category.

Hemifacial atrophy due to morphea (Parry-Romberg syndrome).

KEYWORD: Scleroderma(localized), CREST syndrome, Morphea, Raynaud's phenomenon, Sclerosis of the system

INTRODUCTION:

It is chronic sclerosing connective tissues disease. Its name is derived from the Greek words scleros (hard) and derma (skin). A hidebound skin is important or distinguishing feature the disease, lending credence to term "hidebound disease" [1]. Carlo Curzio of Naples described this disease as a pathological entity in 1752 [2], and Gintrac gave its name, "sclerodermie,". For many years, the disease was considered as dermatological disorder, but when it distinguishing feature of systemic involvement was demonstrated, Goetz coined " progressive systemic sclerosis" in 1945 [3].

It was first described as a pathological entity in the 18th century, but the exact pathogenesis is still unknown due to the disease's rarity. It is recognized that age, gender, genetic, and environmental factors can all influence disease vulnerability [4]. Authors have proposed viral or bacterial infections as the causative agent, particularly in the case of morphea. However, sufficient evidence back up such claiming in lacking [5]. Interaction of immunity with the vascular system, resulting in endothelial damages, has been proposed as a major partner to the disease's initiation and progression [6].

PATHOGENESIS: [7]

*Autoimmune disease: The overproduction of collagen is thought to result from autoimmune dysfunction.

*Genetic, environmental, vascular factors are thought to be causative factors to stimulate the fibroblast.

*Antigens from the human leukocyte antigen [HLA] histocompatibility complex including HLA-B8, HLA-DR5, HLA-DR3, HLA-DR52 and HLA-DQB2 are found to be involved.

VARIOUS TYPES:

Scleroderma is classified into two types:

1. Morphea (circumscribed scleroderma)
   i. Plaque
   ii. Generalised
   iii. Bullous
   iv. Linear
   v. Deep
2. Generalized/progressive scleroderma (diffuse scleroderma)

Morphea scleroderma:

Morphea (localised scleroderma) causes localised fibrosis and is of five forms, but it can also persist in a "mixed form" in which localised scleroderma of face is associated with plaque morphea or linear scleroderma affecting other parts of the body, most commonly trunk. Linear scleroderma, type of morphea, is strongly link to Parry-Romberg syndrome (facial hemiatrophy), but the link is debatable. Linear scleroderma causes unilateral atrophies of the skin, subcutaneous tissues, muscles, and bone structures, as well as neurological deficits [8]. Morphea as previously stated, is classified into five types. In the literature, mixed forms have been reported, as if localised scleroderma of the face with plaque/linear morphea at another region [9].

Generalized/progressive scleroderma (diffuse scleroderma):

Systemic sclerosis (diffuse/generalized/progressive scleroderma) begins as pitting edema of the skin and extends to skin thickening and hardening of the skin. [9] The systemic form can affect multiple organs, including the kidneys, lungs, heart, and gastrointestinal system, with the latter being most commonly involved. Fibrosis or a decreased blood supply both affect these organs. [10]

CLINICAL FEATURES: [11]

*It is characterized by ultimate fixation of the epidermis to the deeper subcutaneous tissues.

*Skin appears yellow, grey or ivory white waxy.

*Skin becomes hardened and atrophic and cannot be wrinkled-described as mask like face and claw like fingers to hands.

*A variant of systemic scleroderma is CREST SYNDROME {C-calcinosis cutis, R-raynouds phenomenon, E-oesophageal dysfunction, T-telangiectasia}

ORAL MANIFESTATION: [12]

*Tongue, soft palate and larynx are common sites.

*Early features include mild oedema followed by induration of mucosal and muscular structures and atrophy.

*Tongue is stiff and board like causing difficulty in eating and speaking.

*Gingival tissue is pale and firm.

*lips appears thin, rigid and partially fixed.

*Dyspagia

*Inability to open and close the mouth.

*Widening of periodontal ligament {2 to 4 times the normal thickness}

*Bilateral bone resorption of angle of mandibular ramus.

*Partial or complete resorption of condyles and coronoid process of mandible.

MANAGEMENT:

There are several autoantibodies that can be used to predict different subtypes of scleroderma. The absence of autoantibodies does’t rule out the diagnosis of the diseases, as 20% of patients with different subtypes of scleroderma do not have these antibodies to regain the immunity. Antinuclear antibodies (ANA) with high specificity for scleroderma include anti-single-stranded, anti-histone, and anti-topoisomerase antibodies. Anticentromere, anti-U3-RNP, anti-Th, anti-fibrillin, antiphospholipid, and antimitochondrial antibodies are also common in scleroderma [13,14].

Treatment for these cases is extremely difficult to diagnose because the exact cause of this disease is unknown. Topical, intralesional, or systemic glucocorticoids, vitamin E, vitamin D3, retinoid, penicillin, griseofulvin, and interferon-alpha are all medical treatments. Though the efficacy of these treatments remains unknown. Restorative plastic surgery (fat or silicone implants, flap/pedicle grafts, or bone implants) is one type of surgical intervention [14,15].

In these cases, a dentist faces a significant challenge in preserving or replacing the existing dentition. Patients instructed on the importance of maintaining proper oral hygiene and counselled on a regular basis to avoid depression caused by the difficult nature of the disease. Mouth dilator exercises, such as putting more tongue blades/ice cream sticks between the back teeth. Sclerodactyly-related toothbrush holding difficulties can be alleviated by advising patients to use adaptive devices such as electric toothbrushes, Waterpik flossers, and floss forks. Muscle relaxants, physiotherapy, and dental appliances can all be used to treat temporomandibular/myofascial pain dysfunction[2]. Dysgeusia and caries caused by xerostomia can be avoided by advising patients to use artificial saliva, sugar-free candies, fluoride toothpaste, and medications such as pilocarpine. [16].

CONCLUSION:

Appropriate scleroderma management necessitates general dentists should monitor patients with scleroderma by performing frequent clinical and radiological examinations to track disease's progression. We have attempted to emphasize the importance of a general diagnosis when diagnosing a rare systemic diseases such as scleroderma, as well as the role of the general dentist in caring for such patients, despite the fact that they are relatively uncommon in general practice.

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