CASE REPORT

AN UNUSUAL PRESENTATION OF A RARE DISORDER – SCLEREDEMA OF BUSCHKE

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ABSTRACT:
Scleredema, also known as Buschke disease (scleredema of Buschke) or scleredema adultorum is a rare connective tissue disorder characterized by progressive thickening and hardening of the skin, usually on the areas of the face, upper back, neck and shoulders. This self-limiting skin disorder is usually associated with conditions like long standing diabetes mellitus with poor glycemic control, recent streptococcal upper respiratory tract infection or paraprotenemia. The pathophysiology of scleredema and its relationship with these associated disorders are not clearly understood. Few therapeutic options are proposed without satisfying results. We report an unusual case of scleredema in a 47-year-old woman who presented with indurated edema of the face, neck and abdomen in the absence of other associated medical conditions.
Key words: Scleredema, Buschke, adultorum.

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INTRODUCTION

Scleredema represents a rare connective tissue disorder of unknown etiology which is characterized by a diffuse, symmetrical, wood-like non pitting induration of the skin with occasional erythema, typically beginning on the face, head or neck and spreading progressively to involve the shoulders, arms, thorax and sometimes extracutaneous sites. [1, 2, 3] Consequently, the range of movement of the affected areas could be affected. Scleredema is often preceded with type 2 diabetes, streptococcal infection, or blood dyscrasias. In case of infectious variant, the onset of the illness usually follows a streptococcal upper respiratory infection. The disorder tends to be self-limiting in majority of cases with the skin infiltration clearing in a few months.[4, 5] Even though skin lesions of Scleredema may resolve spontaneously within months to years, persistent therapy–resistant forms have been described.[4, 5,6] In some instances, especially in cases with visceral involvement the condition may lead to untoward complications.[7-9]

CASE REPORT

A 47-year-old female reported to Department of Oral Medicine & Radiology, Government Dental College, Kottayam, South India with complaints of headache and slurring of speech of 3 months duration. Headache was mainly on the right side which aggravated on mastication. Pain episodes usually lasted for 5-10 minutes. The frequency and intensity of the episodes was noticed to be progressively increasing. Initially she had slurring of speech when she speaks for 5-10 minutes and presently the slurring occurs more frequently, after 2-3 minutes of speech. She had also noticed a thickening of skin over the face, neck, arms and abdomen for the past 3 years which was also progressing gradually. Her past medical history

indicated treated acute glomerulonephritis 20 years back which resolved completely. She also underwent hysterectomy (15 years back) for fibroid uterus and appendectomy 10 years back. She has no history of diabetes mellitus or recent upper respiratory tract infections. Her appetite, sleep and bowel and bladder habits were normal and had a regular menstrual cycle. The vital signs were within normal limits.

Physical examination revealed a woody induration of the skin over the face, neck, shoulder, arms, and abdomen. However, there were no nodules or ulcerations evident over the skin. The neck was short and stiff with difficulty in extending backwards (fig-1).

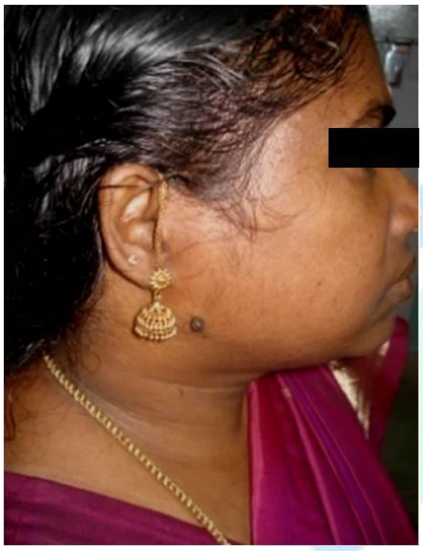

Figure 1: Shows indurated hypopigmented skin over right temple

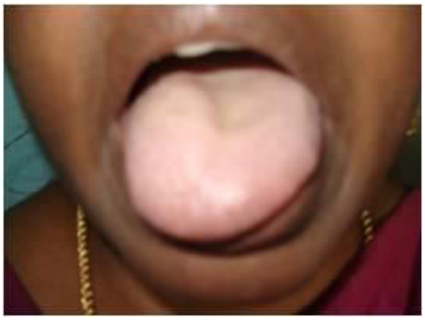

Figure 2: Shows limited tongue protrusion

There was a deviation of angle of the mouth along with decreased protrusive movement of tongue (fig-2). On intraoral examination there was no other significant abnormality except for generalized attrition and early signs of periodontitis. The temporomandibular joint (TMJ) movements were also restricted.

The hemogram, urinalysis, blood calcium estimation, and glucose tolerance test were within normal ranges. Serum analysis for anti nuclear antibody titer and double stranded DNA were negative. Bence-Jones proteinuria was also ruled out. Skin biopsy was preformed and specimen was sent for histopathologic examinations. H&E sections demonstrated thickened dermis with increase in fibroblast and collagen bundles with focal edema. There was mild periappendageal inflammation. Alcian blue staining did not demonstrate mucinous deposits in the dermis. These histopathologic pictures were suggestive of Scleredema. Patient was treated symptomatically and responded satisfactorily and hence the patient is kept under periodic follow up.

DISCUSSION

Scleredema was first described by Piffard[10] in 1876 under the term Scleriasis. It was later modified by Buschke (1902)[8] who renamed it as Scleredema adultorum. In general, Scleredema first affects the face and neck, and then may spread symmetrically to the shoulders, trunk, arms, and legs: however, the hands and feet are usually unaffected. Increased deposition of collagen and mucin (hyaluronic acid, an acid mucopolysaccharide) is seen in this condition; however, the exact pathogenesis is unknown.[4] Women are affected twice as often as men. [9] It differs chiefly from the acute diffuse form of scleroderma by having an excellent prognosis, the skin lesions usually resolving completely. Unlike Scleroderma, the tongue may be involved in scleredema, resulting in dysarthria and difficulty with mastication and tongue protrusion as observed in this case. Scleredema on the face can result in difficulty in opening the eyes and the mouth. [8]

Association of scleredema with DM may be due to accumulation of collagen as the result of irreversible collagen glycosylation which is resistant to collagenase degradation. Additionally, increased collagen synthesis occur due to excess insulin stimulation, microvascular damage, and hypoxia. Theories for nondiabetic scleredema include molecular mimicry, in which streptococcal antigens cross-react with components of the dermis, resulting in an antigen-antibody reaction. This is the possible explanation of scleredema following an upper respiratory infection. [9,10]

Scleredema localized to the back is commonly associated with diabetes mellitus, while generalized scleredema may follow bacterial illness. Comparative quantitative polymerase chain reaction (PCR) analysis of lesional and non lesional skin in patients with scleredema has demonstrated an increase in collagen gene expression in affected sites. [11, 12] Fibroblast culture of affected Scleredema skin has exhibited increased procollagen synthesis. Likewise, experimentally, serum from patients with Scleredema stimulated collagen production in normal skin fibroblasts. Additionally, fibroblasts from the involved skin in non-diabetic patients exhibited biosynthetically activated phenotype, which persists for several years. These alterations are likely to be involved in the development of the cutaneous induration and thickening which is characteristic of this disease. [13]

The morbidity of the skin changes of Scleredema depends on the area of the body involved. The primary lesions of Scleredema are ill-defined, woody, non pitting, indurated areas.[11] Generalized cases may clinically manifest as edema. erythema, hyperpigmentation, and/or a peau d'orange appearance of the affected areas.[7] Scleredema is usually most evident in the upper part of the body, specifically the face, the neck, the trunk, and the upper limbs. The distribution in lower limbs is rare. However, a case of Scleredema affecting the thighs symmetrically has been reported.[7] Involvement of the skin over the joints may cause limited range of motion.

Although rare, extensive truncal involvement may cause restrictive lung disease; one case of fatal scleredema was attributed to pneumonia developing secondary to extensive stiffness of the upper torso.[14] Cardiac involvement in Scleredema patients is rare but may result in myocarditis, cardiomyopathy, heart failure, arrhythmias, pericardial effusion, and unexplained murmurs.[9] Other organs that may be involved in scleredema include skeletal muscles, ocular muscles, the pharynx, the liver, parotid glands, pleurae, the peritoneum, and the spleen. One case report describes Scleredema limited to the periorbital region, which led to partial blindness.[15]

Three clinical groups of Scleredema have been recognized by Graff (Table-1). [11] In the first group (type-1), the disease starts abruptly after an acute upper respiratory tract infection, having a tendency to resolve in a period of months to years; the second group (type-2) begins insidiously without preceding respiratory tract infection and has longer duration over a period of years with paraproteinemia; the third group is of a Scleredema associated with severe long-standing diabetes mellitus (type-3).

Laboratory investigations usually performed in the diagnostic workup of scleredema include:

- Throat culture for group A Streptococcal (GAS) infection and Antistreptolysin O titres to exclude recent GAS infection if you suspect type 1 Scleredema
- Fasting blood glucose and glycosylated hemoglobin (HbA_1C) if you suspect type 3 Scleredema,
- Serum protein electrophoresis and immunoglobulin studies to exclude monoclonal gammopathy, paraproteinemias, including multiple myeloma if you suspect type 2 Scleredema.

Although scleredema has some distinctive clinical features, a biopsy should be performed to confirm the diagnosis. Punch biopsy or incisional biopsy in Scleredema patients should include the subcutaneous fat. The histopathologic analysis reveals a normal epidermis with a thickened dermis and increased spaces between large collagen bundles[9, 11]. The spacing in the dermis results from increased deposition of mucopolysaccharide (hyaluronic acid).[9, 10] The mucin is more prominent in the deep dermis. In some cases, mucin is better detected in unfixed sections stained at a pH of 7.0 with toluidine blue or in tissue fixed with 0.05% or 1% cetylpyridinium chloride solution and stained with Alcian blue at a pH of 2.5. Appendiceal structures in scleredema remain unchanged (unlike scleroderma).

Even though skin lesions of scleredema may resolve spontaneously within months to years, persistent therapy-resistant forms have been described. Appropriate antibiotic therapy should be started in scleredema patients if infection is detected, although antibiotics do not appear to shorten the course of skin findings in scleredema. Various agents have been suggested for the treatment of scleredema adultorum including extracorporeal photopheresis, psoralen and ultra violet A therapy (PUVA), radiotherapy, prostaglandin E 1, corticosteroids and cyclosporine with varying degrees of success.[16-18] Additionally, low-dose methotrexate (MTX) has been reported to be beneficial in scleredema. [1, 3]However, scleredema patients exhibiting diabetes may improve after control of diabetes. Evaluation and treatment for blood dyscrasias and diabetes mellitus should also be completed. Long term follow

up with periodic monitoring of serum protein and immunoprotein electrophoresis to detect the development of paraproteinemia or myeloma or other blood dyscrasias are recommended. The relapse of disease is reported in some cases after apparent improvement.

CONCLUSION

Not many cases of Scleredema have been reported in the literature which indicates the rarity/under reporting of the condition. Importantly, unlike the varieties described in the literature, this case was not associated with any other medical conditions and hence cannot be grouped as any of the known subtypes.

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