SCLEREDEMA- A RARE PRESENTATION

SURESH DHANAPAL
Department of Paediatrics,
MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract:

Scleredema is a disorder of the dermis characterized by brawny edema of the face and neck that spreads rapidly to involve the thorax and arms in a sweater distribution. The involved areas feel indurated and woody, are nonpitting, and are not sharply demarcated from normal skin. In 65-90% of cases, the disease follows an infection. It most commonly seen in female children. Ratio is 21. Most of the time, this scleredema is confused with sclerema, scleroderma subcutaneous fat necrosis. All are separate entity. Unless you do complete physical examination especially palpation, we wont make out the diagnosis of scleredema which is benign condition it has simple treatment complete cure.

Keyword: Scleredema, scleroderma, sclerema, subcutaneous fat necrosis.

INTRODUCTION: Scleredema is an uncommon condition of unknown etiology. It is characterized by a non pitting induration of skin with occasional erythema. It may be associated with history of antecedent febrile illness, diabetes mellitus (27) or blood dyscrasias. Though regarded as a benign, self limiting skin disease, scleredema may be persistent and involve the viscera. Even though it affects all ages, 30% of them occur in less than 10 years. The term scleredema is a misnomer because neither sclerosis nor edema is found on microscopic examination. The histologic findings of scleredema include deposition of mucin (23,25,26) between dermal collagen bundles. The deposition is greatest in the deep dermis.

CASE HISTORY:

4 Years female child first born of non consanguinous marriage brought with history of hardening of skin of 4 months duration and sore throat 4 months back. Hardening was first noticed over face and chest and gradually progressed to both upper extremities over 4 months duration. No history suggestive of systemic sclerosis (scleroderma), hypothyroidism, diabetes & leukemias. If you see in this picture, it seems that she is absolutely normal. But on examination, there was diffuse brawny induration of skin, non pitting & hard in consistency with no limitation of joint & neck.
movements. If you had not done complete examination including palpation, you would have missed the diagnosis of scleredema which is a benign condition. There was no heliotropic rash or gottron’s papules and no raynaud’s phenomenon. Systemic examinations were Negative, ASO titer – raised, ESR - 7 / 20mm (normal limits), Throat swab – no pathogenic organisms, Stool exam -Negative, SS-A,SS-B - Negative. Skin thickness by USG-4mm (1mm is normal in children of same age group). Skin biopsy: Histopathological analysis revealed a normal epidermis with thickened dermis and increased space between large collagen bundles. The space results from deposition of mucin in the dermis. The MPS deposition is more prominent in deep dermis. This specimen slide was fixed with 0.05% cetylpyridinium chloride solution & fixed with alcian blue at pH of 2.5. USG abdomen & chest -Hepatomegaly , no free fluid abdomen, no pleural / pericardial effusion. Thyroid profile : within normal limits. The child was treated with penicillin. On follow up, she improved dramatically after 3 months of penicillin therapy and the skin thickness had decreased considerably which was confirmed by repeating assessment of skin thickness of the same part where earlier one was done with USG. DISCUSSION: Scleredema is an uncommon disease of unknown etiology. It affects both sexes with F:M-2:1 except in the type associated with adult onset diabetes which is more common in men. Though all age groups are affected with scleredema, 30% occurring below 10 years and 50% in less than 20 years age group. Initial skin changes are noticed over face(1,2) and neck with symmetric progression to both sides. Chest, back, shoulder (3) & arms are involved. They are usually confined to the upper part of the body. However, a case of scleredema localized on the thighs has been reported. [8] Hands and feet are typically spared. Scleredema patients with tongue involvement may report with dysarthria. This skin disease is characterised by sudden onset of hardening & induration of dermis as if it is infiltrated with paraffin. There is thickening & hardening of skin, non-pitting, not well demarcated from normal skin as induration fades into normal skin. Facies will be waxy, mask like. Skin feels bound down to underlying skin structures. The overlying skin may be normal in colour, erythematous or hyperpigmented and is not atrophic. Systemic involvement, which is uncommon, is marked by thickening of the tongue; dysarthria; dysphagia [ Tongue, upper esophageal sphincter ]; restriction
of eye and joint movements; and helps to distinguish scleroderma from scleroderma which never affects the tongue. Electrocardiographic changes may also be observed. In 65-90% of cases, it is preceded by streptococcal infections (like sore throat, tonsillitis, pharyngitis, sinusitis, impetigo, cellulitis, mumps, measles, diphtheria, pertussis, typhus fever, scarlet fever, influenza etc. It may be associated with antecedent febrile illness, diabetes mellitus & blood dyscrasias. Though benign, self-limiting skin disease, it may persist & involve viscera. Rarely results in death. Active phase of the disease persists for 2-8 weeks; spontaneous and complete resolution occurs in 6 months to 2 years.

**TYPES:**

Scleroderma can be categorized into 3 clinical subgroups. Each has a different history, course, and prognosis. Note the following:

- **Group 1** includes scleroderma after acute respiratory infection (scleroderma adultorum). Patients in group 1 have a history of a preceding febrile illness, particularly an upper respiratory tract streptococcal infection. The onset of the skin lesions is rapid, and the condition usually clears spontaneously in 6 months to 2 years. The duration is not affected by the use of antibiotics. The term scleroderma adultorum is considered by some to be a misnomer because most pediatric patients fall into this group.

- **Group 2** includes scleroderma patients whose disease tends to occur insidiously, with no preceding illness. This group encompasses cases associated with a monoclonal gammopathy. Group 3 is scleroderma associated with diabetes mellitus (scleroderma diabeticorum) and includes patients with preexisting diabetes, which is typically adult in onset and often type 1. This disorder tends to occur more often in males, and this subgroup of patients typically experiences a more protracted course that is refractory to therapy. As in group 2, the onset of skin lesions is insidious. The upper back typically demonstrates erythema and induration. A pebbled appearance may evolve.

**INVESTIGATIONS:**

- Throat culture to exclude streptococcal infection.
- Antistreptolysin-O titres to rule out recent group A streptococcal infection.
- Fasting blood glucose or glycosylated hemoglobin to R/o diabetes mellitus.
- Serum protein electrophoresis to R/o monoclonal gammopathy. Blood dyscrasias usually appear several years after the onset of scleroderma. (28) Skin biopsy—Epidermis is normal in thickness. Marked rise in thickness (2-3 fold) of dermis with swollen collagen bundles separated by
interfibrous spaces(6). These spaces replaced by acidic MPS in early stage of lesions. Later skin fibrosis may be only finding. In some cases in scleredema patients, mucin is better detected in unfixed sections stained at pH of 7 with toluidine blue or in fixed with 0.05% or 1% cetylpyridinium chloride solution & stained with alcian blue at pH of 2.5. Appendiceal structures in scleredema remain unchanged (unlike scleroderma). In sclerema, skin biopsy specimen will show broadened trabecular fat & space between them will be reduced. Cellular infiltrate is characteristically absent. Fat crystallisation may be seen. Other findings are thinning of epidermis & dermal/subcutaneous tissue fibrosis with edema, thickening of interlobular septae. These are mainly due to reduction in enzymatic desaturation of triglycerides. 6. SS-a & SS-b TREATMENT: There is no specific therapy. 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. Penicillin(24) may be useful. Treat the primary underlying cause. Case reports in scleredema patients also describe scleredema occurring in association with internal malignancies (eg, CA of the gall bladder,(10), malignant insulinoma (11), carcinoid tumor(12). Imaging studies are warranted if this is suggested based on clinical findings.

Rare reports describe scleredema following scabies infestation; appropriate evaluation for possible antecedent infestation may be warranted(13). In 2005, scleredema was reported in association with the use of infliximab; Although rare, treating physicians should consider the possibility of an adverse drug reaction as the underlying etiology(14). No therapy is consistently effective for scleredema. A number of therapies, including systemic steroids, cyclosporine(16,32), methotrexate, high-dose penicillin (15), UVA1 phototherapy(17, E), psoralen with ultraviolet light A (PUVA) either administered systemically or via cream(18,30), or bath therapy(19), penicillamine, electron beam(12,20,29) and glycemic control with prostaglandin E1 (PGE1)(1,31), have all been tried with limited success. In cases associated with myeloma, chemotherapy directed at the hematologic malignancy has been reported to result in concomitant improvement of skin disease(18). For patients with paraproteinemia, extracorporeal photophoresis has been used (21). Myocarditis(7) resulting as a complication from the disease has been successfully treated with penicillin and steroids(7).

DIFFERENTIAL DIAGNOSIS:
1. Morphea,
2. Prolonged systemic sclerosis,
3. Dermatomyositis, mucinosis
4. Scleromyxedema, myxedema
5. Mucopolysaccharidoses – firm, sharply demarcated reddish or purple flat localized solidified area appear over cheek, buttock, thighs. This may calcify later & sometimes heals with atrophic scar,
6. Subcutaneous fat necrosis

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Medicine and Medical Sciences
Sclerema neonatorum Sclerema, scleroderma & sclerema are not same. They are 3 different conditions.

Sclerema neonatorum is mainly seen in newborn babies. It starts from face & legs and it has centripetal progression. It’s not specific for sepsis & also seen in a) Hypothermia b) Gram –ive septicemia c) Hypernatremia. It has got grave prognosis.

COMPLICATIONS: Limited range of motions, poor wound healing, recurrent skin infections, dysarthria dysphagia, difficulty in closing eyes & rarely death can occur.

REFERENCES:
10. Manchanda Y, Das S, Sharma VK, Srivastava DN. Scleredema associated with carcinoma of the gallbladder. British


