SCLEREDEMA WITH SYSTEMIC MANIFESTATIONS

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Summary

A case of Scleredema occurring in old age with systemic manifestations in the form of electrocardiographic abnormalities with a review of literature is presented.

Introduction

Scleredema also commonly known as Scleredema adultorum, first described by Buschke is characterized by sudden onset of stiffening and hardening of the skin and subcutaneous tissue occurring in a symmetrical fashion. Non-pitting diffuse symmetrical induration usually begins on head or neck and spreads rapidly to involve large areas advancing from the neck over to the face and downwards over to the trunk, but usually leaving the hands and always leaving the feet free.

Sometimes there may be associated systemic involvement. The tongue may be swollen causing difficulty in swallowing. There may be restriction of eye movements. Effusion in pleural, pericardial or peritoneal cavity may occur. Electro-cardiographic changes may occur Domonkos[2].

The disease has a tendency to show spontaneous remissions lasting for 6 months to 2 years. Occasionally the disease may last for many decades George et al[3]. No case of scleredema having systemic manifestation has yet been reported from this part of the country.

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Case Report

A seventy year old man was referred from the Medicine to the Skin Department on February 26, 1976. He had one year history of "tightening of skin over the face and chest." He was well about an year ago and noted a rather sudden onset of "tightening of skin" over neck and face which progressed towards the chest. There was no history of sore throat, boils, fever or tonsilitis preceding this illness. There was no history of anginal pain, or myocardial infarction anytime in the past. On examination, patient was found to have "tight and bound down skin over the face, forehead, neck and chest wall". Hands and feet were spared. Skin was diffusely indurated shiny, waxy and cold. The involved skin could not be pinched up. Sensations on the affected area were normal. There was no atrophy or loss of hairs. There was marked puffiness of lower eyelids. Patient had difficulty in opening the mouth. There was no abnormality of the tongue. Clinical examination showed restriction of chest expansion and restriction of the movements of neck and shoulders. Rest of the examination was within normal limits.

Investigations

Urine analysis was normal. Haemogram was within normal limits. Fasting

Blood Sugar was 94 mgm.%. VDRL was non-reactive. L. E. cells were negative on two occassions. The antistreptolysin titre (ASOT) was not done. Skiagram of chest did not reveal any abnormality. Electrocardiogram showed right bundle branch block pattern with auricular rate of 75 per min. and P. R. interval of 0.18 secs. Ventricular rate was 75 per min. Duration of QRS complex was 0.12 secs. Sinus hythm was present.

A skin biopsy specimen from the upper back showed mild atrophy of the epidermis and dense collagenisation in the dermis showing clear spaces between the collagen bundles. There was also mild perivascular infiltration.

Discussion

Although Scleredema was first described by Buschke[1] in 1902, Touraine[4] gives credit to Curzi for describing the first case of Scleredema adultorum in a 17 year old girl in 1775[5]. Piffard in 1876 differentiated this entity from Sceleroderma[6]. Greenburg[3] et al[7] described three cases making the total number of cases described in world literature by that time to 212. Rook[8] described onset of disease in 90-95% cases after an acute febrile illness. Greenburg[7] showed a 65% incidence of preceding infection in this disease. Vallee[9] described two cases of Scleredema who had pleural effusion. One of these had pericardial effusion also. Robinoff and Sweitzer described involvement of tongue in these cases[5]. Vallee[9] described reversible electro-cardiographic changes in Scleredema. Robinoff also noticed electro-cardiographic abnormalities in paediatric cases. Curtis & Shulak[5] noticed reversible electro-cardiographic changes in one case.

In the case presented here the disease started in the late sixties without any preceding streptococcal infection and progressed rapidly from the posterior and lateral aspect of the neck and face to the front and back of chest. There were also electro-cardiographic abnormalities in this case. There has not been any improvement either in the clinical condition or in the electro-cardiographic findings.

REFERENCES

1. Buschke A: Uber Sclerodema, Klin, Wachr, 39; 955, 1902.
2. Domonkos AN: Andrew's Diseases of the Skin, 6th edition, 198, 1971.
3. George T, Fernandes R and Dhurandhar MW, et al: Scleredema, A case report and review of literature, Ind J Derm Vener, 41: 66, 1975.
4. Touraine A, Gole L and Soulignac R: La cellulite Scleroderimiforme Extensive Benigne, Ann Derm Syph. 8: 767, 1937.
5. Curtis AC and Shulak BN: Scleredema adultorum, Arch Derm, 92: 526, 1965.
6. Saffron MH: Scleriasis (Scleredema adultorum, as first described by Piffard, Arch Derm Syph, 47: 110, 1943.
7. Green Burg LM, Seffert TC and Worthen HG et al: Scleredema adultorum in children. Ped. 50: 261, 1963.
8. Rook A: Scleredema, Post-Grad Med J, 30: 30, 1954.
9. Vallee BL: Scleredema-A systemic disease, New Eng J Med, 235: 207. 1946.