Scleredema Adultorum Of Buschke: A Case With Multiple Causes

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Citation

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Abstract

Background: Scleredema adultorum of Buschke is an uncommon skin condition that may accompany following diabetes mellitus, proceeding of infection or may be associated with monoclonal gammopathy,. Our case had it with multiple other underlying causes.

Observation: This is report of a case with scleredema of Buschke with history of eight months. The patient had no history of a definite diabetes mellitus, but only an impaired glucose tolerance test. Hyper IgG gammopathy with increasing of ASO titer in association with a history of preceding chronic infection of both legs following accidental fractures since of two years ago were seen also. Histological findings of a biopsy specimen from involved neck skin showed marked thickening of the reticular dermis and collagen fibers has been broaden and separated by clear spaces which showed positive reaction with alcian blue. Anti streptolysin O titer in his serum level was more than 1000 UI/ml (international union / per milliliter). Glucose tolerance test (2 hours after oral eating 70 gram anhydrous glucose powder) showed impaired (>200mg/dL) in three times doing exam, but glycosylated hemoglobin A-1c was as 4.5 as in goal limitation (action suggested >6.4).

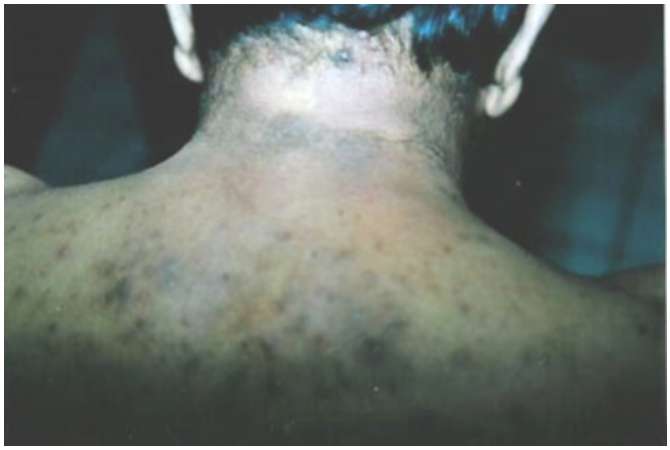
Results: Therapeutic plan included oral penicillin and cyclosporine and azathioprin that were not significantly effective during the last 7 months, but the patient feels mild improvement in the last month.

Conclusion: This is an uncommon case of progressive scleredema adultorum of Buschke in a man with multiple underlying causes with mild impaired glucose tolerance test, increased G type of immunoglobulin (IgG) of 1942 mg/dl (normal range =700-1600mg/dl), and increased serum level of ASO titer, as well as history of chronic over imposing infections on both leg fractures and after operations.

CASE REPORT

A 32-year old man is referred to dermatology clinic due to bilaterally hardening of neck distributed to both shoulders following of appear of some sparse papules and pustules on the surface of skin. He noted hardening and tightness of the skin firstly over his posterior neck and upper back area approximately 3 moths before referring to this center. Woody sclerosing edema of upper proximal limbs which has been appeared at these site and progressed to anterior chest wall and symmetrically on both arms, These findings in our case were typically in favor of scleredema adultorum (Buschke type) (Fig 1).

Figure 1

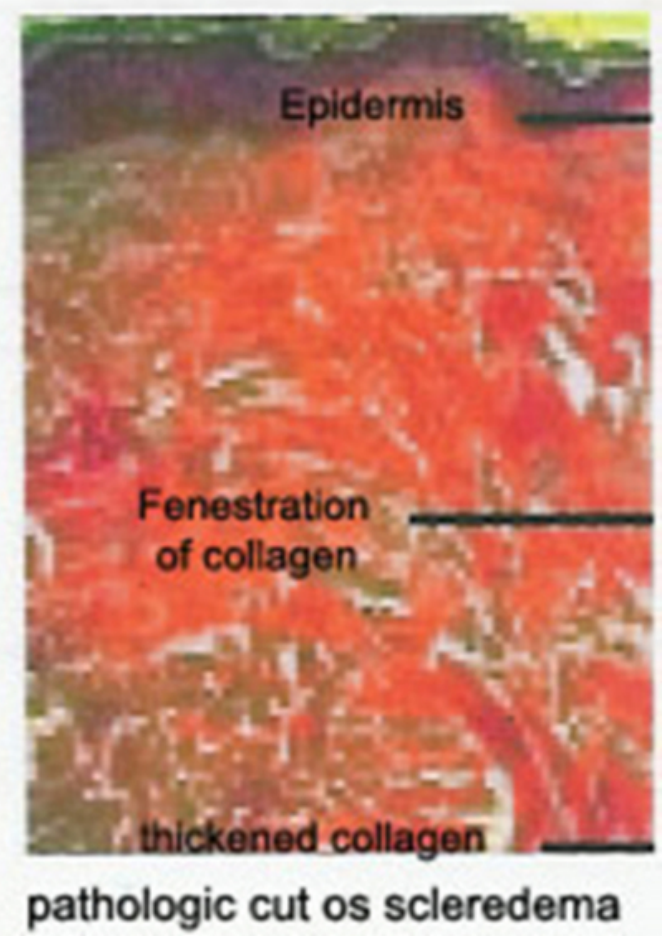


He has been admitted in dermatology field of a general hospital for ruling out of cause of scleredema adultorum of

Buschke. Physical examination showed woody skin over the posterior surface of neck and upper back, which was thicker and more firm than normal in touching. Some papules and pustules are seen on neck surface. Involvement in the first two months was in association with mild limitation in laughing and facial figuration but has not any problem in breathing, swallowing, and in eating, or eye movement, but he suffered of uncomfortable sight seeing around.

In the past history he mentioned fractures of both legs following a traffic accident 2 years ago. He has used many different drugs such as antibiotics and non steroidal anti-inflammatory drugs because of superimposing infection in the fracture area. Leg fractures have been fixed by 3 plates two years ago, but two of them have been extracted 6 months ago, and only one remains now. A 4 mm deep punch skin biopsy was obtained at the neck area, and stained in Hematoxylin and Eosin. Sections of thick reticular dermis with thick collagen bundles separated by clear spaces (fenestrations) are seen. Basophilic myxoid materials were found focally and were present within some of these spaces whom were compatible with scleredema adultorum (Fig 2).

Figure 2



Laboratory data contains complete blood count, fast blood glucose (FBS), rheumatoid factor, thyroid hormone serum level, creatinine, and blood urea nitrogen, all were in normal ranges. Glucose tolerance test showed disturbance, so FBS was 80 mg/dl, but, 1,2 and 3 hours later after using glucose powder orally, serum glucose showed as 186, 225 and 171 mg/dl respectively in the three times testing was repeated. Glycosylated hemoglobin (A1c hemoglobin) of serum was at goal limitation (usually action suggested range is >6.4). A serum protein electrophoresis showed total protein = 8.6 (normal was 8gr/dl) serum albumin = 4 and mild elevated gamma immunoglobulin 1942 mg/dl (normal range is 700 to 1600 mg/dl). Therapeutic plan in him was contain oral penicillin and cyclosporine that were not significantly effective during last 7 moths but stiffness has been reduced mildly at last month which the patient was on taken of (piascledin)R capsules and azathioprin (Immuran)R (50mg tablets) (150mg/daily).

DISCUSSION

Scleredema adultorum is a connective tissue disorder characterized by progressive symmetric indurations and

thickening of the skin, principally over the posterior neck and upper back skin ([1]). Monoclonal gammopathy was observed in three patients with long-term and two cases more next with widespread scleredema (Buschke's disease) ([2,3]). Beginning of the disease in our case was at the neck and back, although the etiology of scleredema is unknown. Increased deposition of collagen fiber and mucin compounds following gammopathy are seen also in affected area ([4]). In addition to the associations with diabetes mellitus, infection, paraproteinemias, and multiple myeloma, has been mentioned ([5,6,7]). Our case had gammopathy and multiple other underlying causes contain: a: a past history of chronic infection during last1.5 years after traumatic fractures and also following operation b: high level of anti streptolysin O serum level (ASO) titer, c: increasing of IgG serum level and d: having an abnormal glucose tolerance test. One-fourth of scleredema cases are abrupt in onset and occur shortly after an infection, usually streptococcal pharyngitis. He denied any history of pharyngitis and we think scleredema in our case can be related to every one or related to both or all of mentioned multiple underlying causes. He had a history of chronic leg infections following fractures, and history of other infections have been reported in association with acute-onset scleredema include influenza, measles, mumps, and varicella. He denied any history of similar diseases in last 5 years.

Scleredema tends to present in young than aged and in women cases against men as 2 over 1. The lesions usually begin on the face and/or neck and spread to the arms, shoulders, back, and chest. In the present case progression of disease was different, and mask like expression has been developed from neck and back. He had no complications such as dysphagia, breathing or cardiac complication or arrhythmias. There is no clear demarcation between involved and normal skin. Although he has not showed any sign of improving during last 7 months therapy, mild improvement has been found during last month. The disease is usually resistant to therapy but approximately 50 percent of the cases with infection-associated are self-limited and resolve over months to years ([1]). This is an uncommon case of progressive scleredema adultorum of Buschke in a man with multiple underlying causes.

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