Case 3 : Scleredema

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CASE SUMMARY

History

Mr. Lau is a 50-year old ethnic Chinese male. He complained of 6 months' history of asymptomatic thickened skin plaque over the back of neck, upper back and bilateral shoulders. There was no Raynaud's phenomenon, dysphagia, or joint pain. There was no preceding history of upper respiratory tract infection. He had hypertension for more than a year which was well controlled with Renitec and Betaloc. There was no diabetes or ischaemic heart disease. The family history was unremarkable.

Physical examination

There was symmetrical, ill-defined, skin-coloured, indurated plaque involving the posterior aspect of neck, upper back and both shoulders. (Figure 1). Face and acral sites were not involved. There was no muscle weakness or tenderness.

Differential diagnoses

The following diagnoses were considered: scleredema, scleredema (localized), scleromyxedema, normal variant.

Investigations

The following blood tests, namely complete blood count, erythrocyte sedimentation rate, liver/renal function tests, fasting blood sugar, ASOT titre, thyroid function test, immunoglobulin pattern, and Rheumatoid factor, were done. The results were all within normal limits.

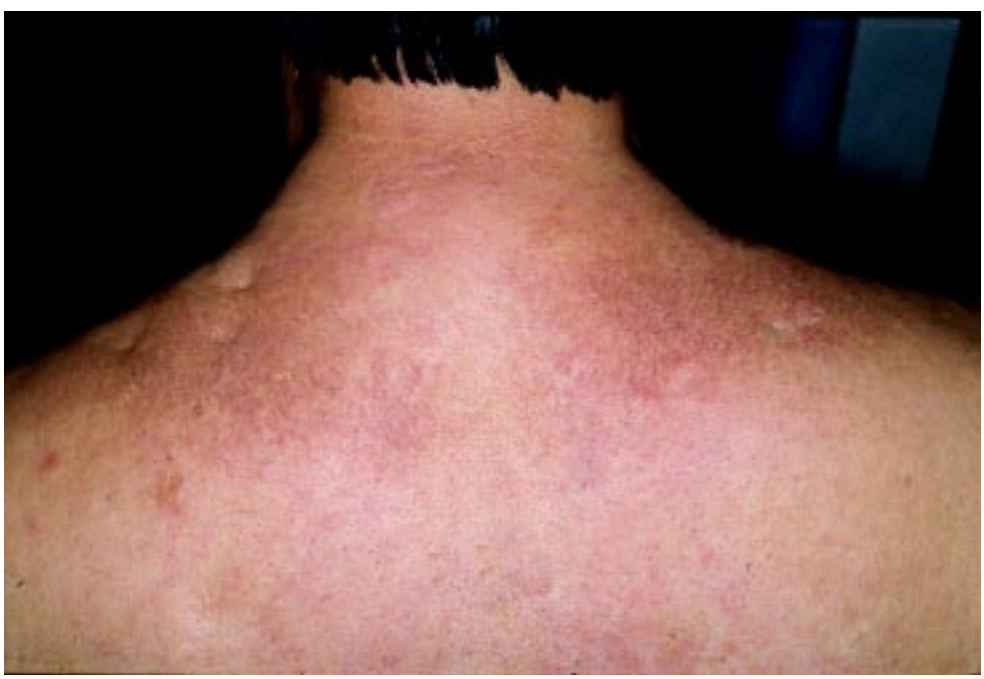

Figure 1: Symmetrical, ill-defined, skin coloured, indurated plaque involving the posterior aspect of neck, upper back and both shoulders.

Skin Biopsy was performed which showed unremarkable epidermis and the dermis was greatly thickened. The eccrine glands were abnormally situated within the upper third and mid dermis. The collagen fibres were broadened and separated by mucin. Mast cells were increased but the fibroblasts were not increased. There was mild superficial perivascular chronic inflammatory cell infiltrate in the dermis. The features were compatible with Scleredema of Buschke.

Management

Reassurance was given to the patient. Regular follow up was necessary to monitor progress. The possible association with other medical conditions such as cardiac involvement and paraproteinaemia should be ruled out.

Discussion

In scleredema, the histology is characterised by homogenisation of collagen fibres which is less pink in colour. Scleromyxedema is characterised by increase in the number of fibroblasts.

A local survey of scleredema from Social Hygiene Service had collected twelve cases. Eighty-three percent were associated with diabetes. Among these patients, 50% were insulin dependent. Most of them had hypertension. It is worthwhile to rule out diabetes by fasting blood sugar or oral glucose tolerance test.

REVIEW ON SCLEREDEMA

Scleredema is a rare, idiopathic, connective tissue disease characterised by non-pitting swelling and induration of the skin. It was first recognised by Buschke in 1902 as a distinct entity. Previous synonyms (scleredema adultorum, scleredema neonatorum) were dropped due to frequent occurrence in children. In a review of 209 cases, 29% occurs in patients younger than 10 years of age, 22% at the age of 10-20 years, 49% in adults.

Aetiology

The aetiology is unknown. There were various proposed, but unsubstantiated, hypotheses such as streptococcal hypersensitivity; lymphatic injury; disorders of the peripheral nervous system or pituitary function.

Histochemical analysis[1]

There were increased amounts of glycosaminoglycan (GAG) and collagen in affected skins. Fibroblasts from the affected skin produced increased amounts of GAG in vitro than those from uninvolved skin. There was markedly increased type I collagen expression. Serum from patient had been shown to stimulate biosynthesis of collagen by normal or autologous skin fibroblasts.

Clinical features

There are three clinical forms: abrupt onset; insidious onset; association with severe complicated diabetes[2].

Abrupt onset

It is preceded by viral or bacterial febrile infection, such as streptococcal infection, influenza, measles and mumps, a few days to 6 weeks before the onset of skin lesion. The prodromal symptoms include fever, malaise, myalgia and arthralgia. There is marked, non-pitting, symmetric skin induration with wood-like consistency. The lesion has a waxy white discolouration or shiny in appearance. The border is ill-defined which gradually transformed from involved to normal skin. The common sites of involvement include the posterior and lateral aspects of neck, face, shoulder and upper trunk. Skin over joints can be involved resulting in restriction in movement. Facial involvement causes expressionless facies, difficulty in wrinkling and smiling. Tongue and pharynx may also be affected causing dysphagia. This subtype may clear within a few months or may take a prolonged course.

Insidious onset

There is no definite preceding illness. It spreads gradually and runs a chronic course.

Associated with severe complicated diabetes

It has an insidious onset. The skin induration may be preceded by erythema or pustules. The condition persists indefinitely. The associated diabetes is of maturity-onset, insulin requiring and is difficult to control. Patients are usually obese and have complications of diabetes like retinopathy, nephropathy, neuropathy, ischaemic heart disease, peripheral occlusive arteriopathy.

The possible pathogenic role of diabetes was the altered mucopolysaccharide metabolism, susceptibility to infectious agents, or accelerated collagen aging. Antidiabetic therapy has no effect on the evolution of scleredema.

Other reported associated conditions

A number of conditions were reported in association with scleredema. Monoclonal gammopathy was one of the association, usually IgG para-proteinaemia. But IgA and IgM paraproteinaemia had also been reported. The light chains involved could be the l or k type. Cardiac abnormalities reported include diastolic gallop without evidence of heart failure in children, and repolarisation abnormality(S-T depression, T inversion) in ECG. Rheumatoid arthritis, Sjogren's syndrome, primary hyperparathyroidism, and malignant insulinoma were also reported.

Pathology

The epidermis is normal. The dermis is three times thicker than normal. The collagen bundles are thickened and separated by clear spaces which are referred as 'fenestration'. The eccrine ducts are located abnormally high in the upper or mid dermis, due to the replacement of subcutaneous fat by dense collagenous bundles. There is no increase in the number of fibroblasts. There is mild superficial perivascular infiltrate. Increase in mast cells is present. The ground substances in the fenestration are stained metachromatically with toluidine blue, owing to the presence of hyaluronic acid.

Differential diagnoses

The differential diagnoses include scleredema, morphoea, scleromyxedema, dermatomyositis.

Treatment

There is no effective treatment.

Learning points:
In Scleredema, it is worthwhile to rule out diabetes by fasting blood sugar or oral glucose tolerance test.

References

1. Varga J, Gotta S, Li L, et al. Scleredema adultorum: case report and demonstration of abnormal expression of extracellular matrix genes in skin fibroblasts in vivo and in vitro. Br J Dermatol 1995;132:992-999.
2. Venencie PY, Powell FC, Su WPD, et al. Scleredema: A review of thirty-three cases. J Am Acad Dermatol 1984;11:128-34.