ABSTRACT

Aim: To recognize a line of treatment for scleredema of Buschke in an adolescent.

Background: Scleredema of Buschke is an uncommon disorder characterized by induration of the skin, which includes a non-pitting hardening of the skin around the neck, shoulders, and trunk sometimes the face. Three variants are recognized. The histopathologic features of scleredema are characterized by thickened collagen bundles within the reticular dermis that are separated by mucin containing fenestrations. No consistent treatment modality is currently followed.

Case report: The present case report describes scleredema of Buschke in a 10-year-old female child reported with stiffness of facial skin and difficulty in opening the mouth. The patient was treated with antibiotics and vitamin supplements and there was drastic improvement with decrease in skin stiffness and increase in mouth opening. The stiffness of facial muscles decreased and mouth opening increased.

Conclusion: Multiple treatment modalities for scleredema have been used. In the present case, high-dose antibiotic therapy was used with good prognosis.

Clinical significance: High-dose antibiotic therapy can be used for treatment of dermatologic disorders with reasonably good prognosis.

Keywords: Scleredema of Buschke, Sjogren’s syndrome, Penicillin.


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Conflict of interest: None declared

BACKGROUND

Scleredema of Buschke is an uncommon disorder characterized by induration of the skin that includes a non-pitting hardening of the skin around the neck, shoulders and trunk, thickening usually begins on the neck and later spreads to the shoulders, upper part of the trunk and sometimes the face.¹²
examination of soft tissue, loss of stippling seen in gingiva, buccal mucosa was firm in consistency. Hematological investigation showed that the blood counts, blood sugar, blood urea, serum creatinine were within normal range and ASO titer >200 IU/ml.

Histopathological examination of a skin biopsy from the neck and buccal mucosa showed (Figs 4 to 6) swelling and splitting of dermal collagen bundles by an increase in ground substance, dermis was 3 times its normal thickness, deposition of hyaluronic acid between the collagen fibers, in dermis thick collagen bundles separated by large interfibrous spaces, fibrous noncellular hyperplasia replaces large areas of subcutaneous tissue resulting in increased skin thickness, increase in acid mucopolysaccharides.

The patient was treated with antibiotics and vitamin supplements and there was drastic improvement with decrease in skin stiffness and increase in mouth opening (Table 1 and Fig. 3) in her follow-up visits.

**DISCUSSION**

Buschke in 1902 described this systemic disease with woody, nonpitting indurations of ‘plaster statue’ consistency starting at the nape of the neck and spreading throughout the body, usually symmetrically but sparing the palms and soles, rarely the tongue, pharynx and parotid gland may also be involved, there is a history of preceding infection in 65 to 90% of cases as reported in

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<th>Table 1: Measurements showing mouth opening</th>
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<td>1st visit</td>
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the present case, however, associations also have been reported with diabetes,\textsuperscript{9} monoclonal gammopathy,\textsuperscript{10} multiple myeloma,\textsuperscript{11,12} primary hyperparathyroidism,\textsuperscript{13} rheumatoid arthritis\textsuperscript{9} and rheumatoid arthritis with sjogren syndrome.\textsuperscript{14}

The condition is usually self-limiting with active phase persisting for 2 to 8 weeks followed by spontaneous and complete resolution in 6 months to 2 years. Occasional cases with no resolution even after 10 years have been cited. The potential chronicity and severity of the disorder should not be underestimated, as there have been at least two deaths due to scleredema.\textsuperscript{15,16}

Histologically, scleredema shows a normal epidermis but the dermis may be three times the normal thickness with swelling and splitting of the dermal collagen bundles and variable deposition of the mucin in the dermis, histochemical analysis have shown that the amount of glycosaminoglycans and collagenase increased in the affected skin.

**CONCLUSION**

There is no definitive therapy for scleredema. In the mildest cases, optimization of metabolic control and physiotherapy especially in case of motion or respiratory disability are recommended. For the rare progressing forms, various systemic treatments have been proposed including corticosteroids, D-penicillamine, cyclosporine, methotrexate, PUVA therapy, prostaglandin E1, factor XIII or radiotherapy. High-dose antibiotic therapy can prove to be a useful line of treatment of this rare disorder.

**CLINICAL SIGNIFICANCE**

Oral symptoms of scleredema are often neglected. Patients with this disease may have difficulty in mouth opening. High-dose antibiotic therapy and vitamins may be used for treatment of scleredema with reasonably good prognosis.

**REFERENCES**


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