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Scleredema adultorum of Buschke in a child – a rare case

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ABSTRACT

Scleredema adultorum of Buschke is a rare connective tissue disorder reported usually following streptococcal infection, influenza, measles, and mumps. This is a report of scleredema adultorum developing after streptococcal infection in an 10-year-old girl. The diagnosis was established by characteristic picture on skin biopsy using special stain. The patient had a benign course and responded to local emollients only.

Key Words : scleredema, child, skin biopsy, ASO titre

INTRODUCTION

The scleredema Buschke belongs to the group of mucinoses, rare connective tissue disease characterized by hardening of the skin, symmetrical, insidious onset, with a predilection for the upper regions of the posterior trunk, neck and shoulders. Extracutaneous manifestations of the disease has been also described.[1] Though the benign nature of the disease is the rule in most cases, the systemic involvement has to be looked for and appropriately treated to prevent morbidity and mortality in scleredema adultorum of Buschke. Here we report a case of scleroderma in a child of 10 years.

CASE REPORT

A 10 year old girl presented with progressive thickening of the skin starting from the face and gradually progressing downward symmetrically to involve trunk (both chest and back), upper arm and proximal part of forearm over 3-4 weeks. There was difficulty in opening the mouth with masked facies. Child was afebrile without any feature of dysphagia, arthralgia, dyspnea, rash or raynaud's phenomenon. Child was non diabetic. She had history of fever with sore throat few weeks prior to the appearance of symptom, which was subsided by antibiotics. There was no such family history. Systemic examination was normal except skin over trunk and face was hard, non pitting, non tender. The area could not be pinched. There was obliteration of nasolabial folds and forehead wrinkles. There was no restriction of chest expansion. Trunk and extremities were involved symmetrically. Skin over hand and finger tips was normal.

We investigated the child with complete hemogram, ESR, albumin/globulin, sugar/urea/creatinine, mantoux, ECG, CXR,CRP, RA factor, ANA. Those tests were normal except for ASO titre which was significantly raised(> 200 iu/dl). Skin biopsy was taken from deltoid region which showed normal epidermis with thick dermis with swollen collagen bundles separated by an increase in ground substance with normal adnexal structure. An alcian blue staining revealed mucin

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deposits between the collagen bundles(Fig-1). These findings were compatible with sclererderma. We treated the child with local emollients. She started showing improvement over few weeks with tendency towards gradual softening of skin symmetrically especially over extremities with appearance of forehead wrinkles and nasolabial grooves.

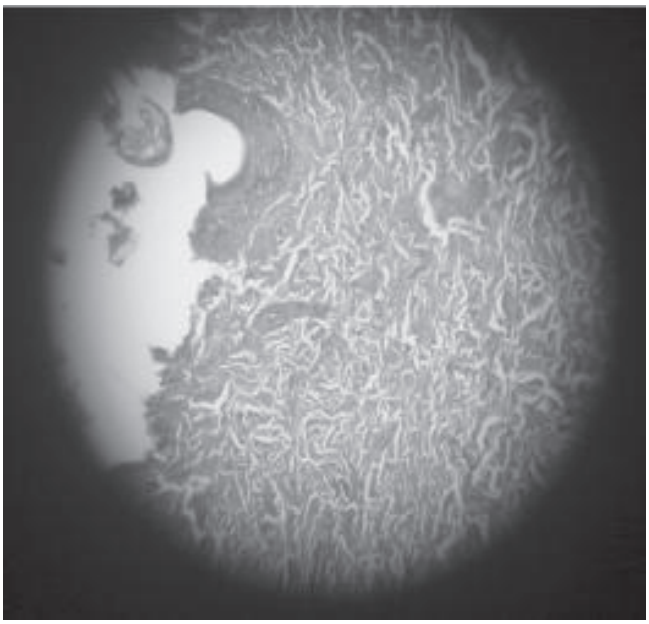

Figure 1: Mucinous deposits between the collagen fibres of the dermis in alcian blue staining of skin biopsy (H& E,100X)

DISCUSSION

Sclererderma is a rare connective tissue disease with increased amounts of collagen and glycosaminoglycans.[2] It is a scleroderma-like disorder, which is frequently misdiagnosed as systemic sclerosis. It can be differentiated from Systemic sclerosis, Morphea and Myxedema by skin involvement clinically, usually lack of systemic features, scleroderma specific autoantibodies and Raynaud's phenomenon.[1] It is classified into 3 different types. Type 1 scleredema is referred to as adultorum of Buschke since the post-infectious form of scleredema was the one, first described by Buschke himself. Type 1 consists of predominantly paediatric patients with acute onset preceded by a febrile illness, particularly streptococcal infection. Viral infections like influenza, cytomegalovirus, measles, pertussis, mumps and diphtheria are also reported. A case following chickenpox has also been reported.[4] Type 1 has benign course , excellent prognosis and spontaneous resolution over months. Other two types are mainly found in adults and mostly associated with paraproteinemia, multiple myeloma, rheumatoid arthritis (Type 2) or type 2 diabetes mellitus (Type 3).[2,3,5] They are mostly insidious in onset and difficult to treat. In majority the disorder is restricted to skin. Tongue involvement is reported. Extracutaneous involvement, including oesophagus, heart, pleurae, skeletal muscles and eyes with periorbital oedema, restricted motility and sicca syndrome have been reported rarely.[1,4] Recurrent attacks are unusual. Histopathological examination of skin biopsy in scleredema shows thickening of dermis with deposition of mucin and collagen with maintained ductal structures.[6] The treatment protocol for scleredema adultorum has not been effectively described in the literature. Steroids and immunomodulators have been tried for systemic involvement of this disease.[7]

CONCLUSION

Scleredema, though very rare in children should be kept in mind whenever child presents with thickening of skin and treated accordingly.

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