CUTIS LAXA

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A 20-year-old male had cutis laxa manifesting as loose pendulous skin over the face and neck giving a prematurely aged appearance.

Key word : Cutis laxa.

Cutis laxa, also called dermatochalasis, generalised elastolysis, chalazoderma, as the name suggests, is characterised by lax, loose and pendulous skin, resulting in a prematurely aged look. The changes occur in the skin all over the body, but rarely it may be localised. The pathogenesis is not exactly known, though deficiency of elastase inhibitor has been postulated.

Case Report

A 20-year-old male farmer, born to non-consanguinous parents was seen for loosely hanging and pendulous skin over the face and neck, of 10 years duration. The patient had a prematurely old look, causing him a cosmetic problem. There was no preceding dermatoses, drug intake, dyspnoea, palpitations, urinary or gastro-intestinal symptoms.

Clinical examination revealed a well-built individual with no signs of systemic involvement. The skin over his face and neck showed gross laxity and had a doughy feel (Fig. 1). The skin could easily be pulled away from the underlying tissues and when released returned very slowly to its former position. The skin on other parts of the body was normal.

Histopathology of the facial skin with Verhoef's stain showed epidermis and dermis of normal thickness. The elastic fibres of the dermis were decreased and showed granular degeneration while the collagen appeared normal. Skiagram of chest, electrocardiogram, ECHO, Doppler studies and barium meal studies were all normal. The patient was referred to the plastic surgeon who decided to perform rhitidectomy as a face lift surgery.

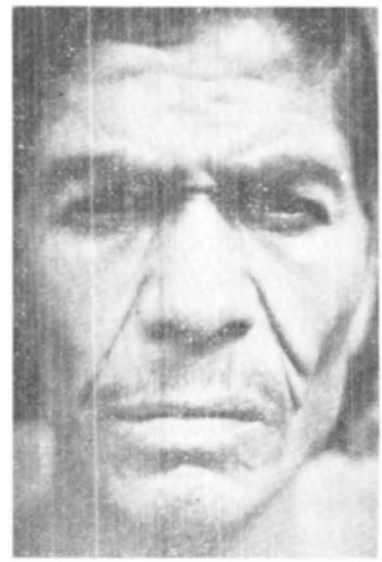

Fig. 1. Loosely hanging pendulous skin over the face.

Comments

Cutis laxa is of two types—congenital and acquired. In the congenital type, the usual mode of inheritance is autosomal recessive, although autosomal dominant transmission has been described.[1] In the acquired type which has no genetic background cutaneous changes usually appear at or soon after puberty, but may appear in childhood, as in our case, or not until middle or old age.

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In both the congenital and the acquired types, due to a defect in the elastic fibres, internal organs are frequently involved, and manifest as pulmonary emphysema, pulmonary valvular stenosis, hernia and diverticula of the gastro-intestinal tract.[2,3] None of these changes occurred in this patient.

Histopathology of skin needs special stains for elastic tissue, to reveal a reduction in elastic fibres in the early stages, while in the late stages the elastic fibres are replaced by fine dust-like granules scattered in the dermis.[1,5]

The disease must not be confused with Ehlers-Danlos syndrome, where the skin is hyper-extensible but not lax. In pseudoxanthoma elasticum the skin may be lax at the sides of the neck, but is characteristically yellow and the face is usually spared. The folded loose skin in leprechaunism is thickened and not lax.

Cutis laxa can be effectively treated only by plastic surgery to reduce the cosmetic disfigurement. All cases should be followed-up for any systemic involvement.

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