Case Report: Acquired Cutis Laxa in a Six Year Old Girl

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ABSTRACT

Cutis laxa is a rare skin disease of unknown aetiology. Evaluation of the skin shows loss of elastic fibers with resultant laxity of the skin. We present the case of a 6 year old girl who had normal skin until age 2. She presented with a four year history of generalized pruritus, lax skin and looked older than her age.

KEYWORDS: Acquired Cutis Laxa, Generalised Pruritus, Lax skin.

INTRODUCTION

Cutis laxa (CL) is a rare skin disease of unknown aetiology with defects in elastic tissue synthesis or its destruction.[1-3] Cutis laxa can be congenital or acquired,[4-7] characterized by loose inelastic skin. Acquired cutis laxa (ACL) is rare, occurs in both children and adults,[2,8] and can be generalized or localized.[6,9,10]

CASE REPORT

A 6 year old girl presented to us with a 4 year history of loose skin and generalized increased skin darkening. At age 2, she developed generalized pruritus followed by asymptomatic progressive skin darkening and development of folds in the flexures. There was no history of rash, insect bites or popular urticaria. There was also no history of atopy, drug allergies or similar skin disease in the family or in their neighborhood. Ante natal history and developmental milestones where normal. The girl was not a product of a consanguineous marriage.

Physical examination revealed a normal sized 6 year old looking older than her age. Her skin was hyperpigmented, velvety with generalized creases. The skin around her neck and axillae hung in redundant folds (Fig. 1&2). There was no evidence of localized inflammatory disease. She emitted a peculiar smell. Systemic examination was normal.

Biopsy from the lax skin of the armpit showed paucity of elastic fibers in the dermis (Fig 4). No inflammatory cells were seen in the dermis. Other investigations were normal. A diagnosis of acquired cutis laxa was made.

DISCUSSION

Clinically, CL manifests as lax skin and a premature aged appearance which may involve the entire skin surface especially the face, neck, back and thighs.[1,3,10] The skin hangs in redundant folds especially in the neck and axillae as is the case with our patient.[10-12] Acquired cutis laxa can be idiopathic[9,10,13] or associated with other conditions (Table 1).[2,6-8,10,12-21]

Histology of the affected area often shows reduction or loss of elastic tissue in the dermis.[14-1] The cause of acquired cutis laxa is unknown. However, the presence of infiltrates at the acute stages of the illness suggests that, the chemical mediators released from the cells may be responsible for the eventual elastolysis seen in the skin[3]

In African children, 18 cases of post-inflammatory elastolysis and cutis laxa (PECL) have been described. In most of the children, it occurred after obvious inciting conditions.[18] It followed recurrent acute inflammatory lesions in previously healthy female children aged one to four years with no family history of similar illness and no systemic

involvement as in our patient.[18]

Although our patient did not have any rash, she had generalized pruritus suggestive of an ongoing pathology at the onset. There were no investigations at the onset to show if she had eosinophilia as documented by the earlier cases seen in Kenya and South Africa.[18] She had no eosinophilia at presentation to the clinic, no systemic involvement and her skin biopsy showed no inflammatory cells typical of the chronic phase seen in the previous cases of PECL.

The aetiology of PECL is unknown but an allergic reaction to an unknown inciting factor such as arthropod bites has been suggested especially in those with localized lesions at the onset.[2,18] The rarity of this condition in the tropical environment would suggest other rarer inciting agents than arthropod bites or a multifactorial aetiology requiring insect bites and possible genetic predisposition not explored in our patient.

We conclude that our patient most likely had an initial inflammatory disorder which led to the cutis laxa. It is possible that this is a variant of what was initially called PECL in earlier literature.[2,18] The rarity of acquired cutis laxa suggest other factors besides environmental in its aetiology.

To the authors' knowledge, this is the first reported case of acquired cutis laxa in a Nigerian child following an inflammatory skin condition.

AUTHORS	ASSOCIATION
Fontenelle E et al[2]	Neutrophilic dermatosis
Peters T et al[6]	Inflammatory arthritis
Hoang MV et al[7]	Cutaneous mastocytosis
Yadaw TA et al [8]	Multiple myeloma
MitraS et al[10]	Itchy skin
Kumar P et al[12]	Itchy rashes
Wang S[13]	Papular lesions
Lee MY et al[14]	Multiple myeloma and Systemic amyloidosis
Mahajan VK[15]	Cutaneous mastocytosis
Golisch KB et al[16]	Urticarial vasculitis, Systemic lupus erythematosus
Majithia RA et al[17]	Light and heavy chain deposition disease
Verhagen AR et al [18]	Post inflammatory
Koch SE et al[19]	Drugs (Isoniazid)
BharadwajS et al[20]	Recurrent ileus
Nirmal B et al[21]	Inflammatory Bowel Disease, Inflammatory Arthritis and IgA Nephropathy.

Table 1: Associations of Acquired Cutis Laxa

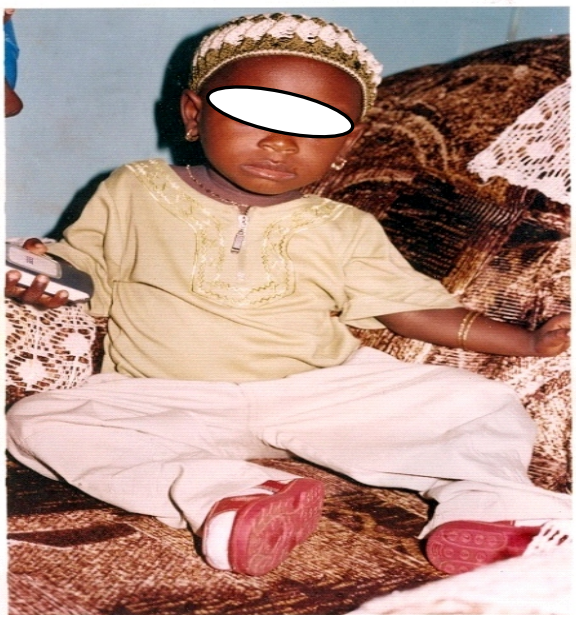

Figure 1a: At age 9 months

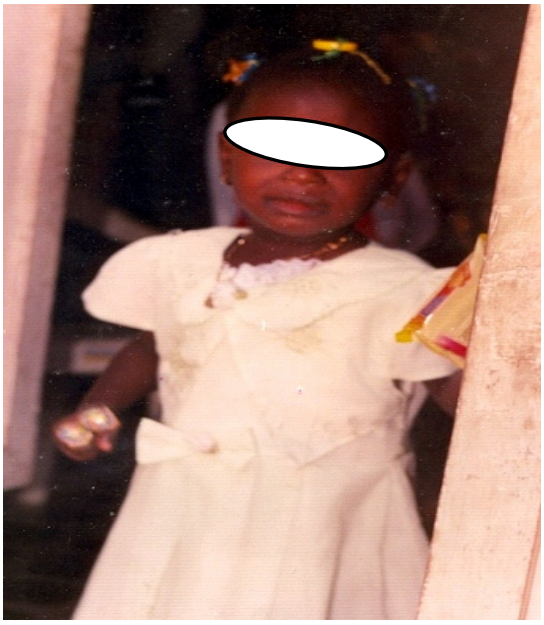

Figure 1b: At age 2 years

Figure 2: Redundant Skin Appearing in Folds in the Neck

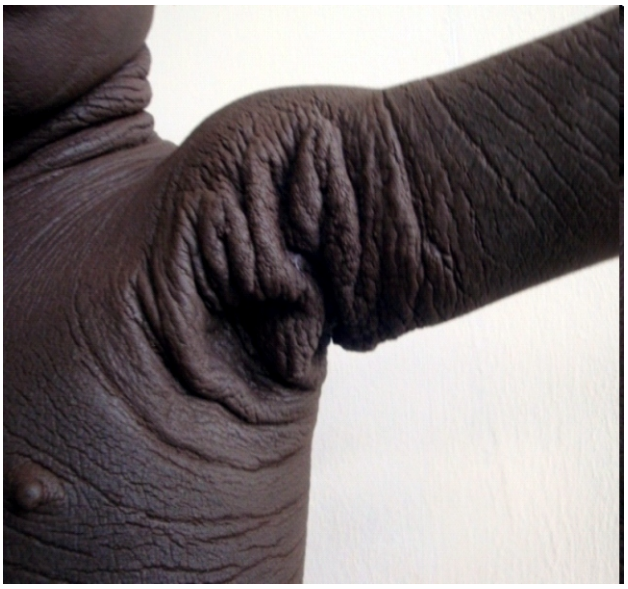

Figure 3. Skin in axilla hanging in folds. Surrounding skin is also thickened

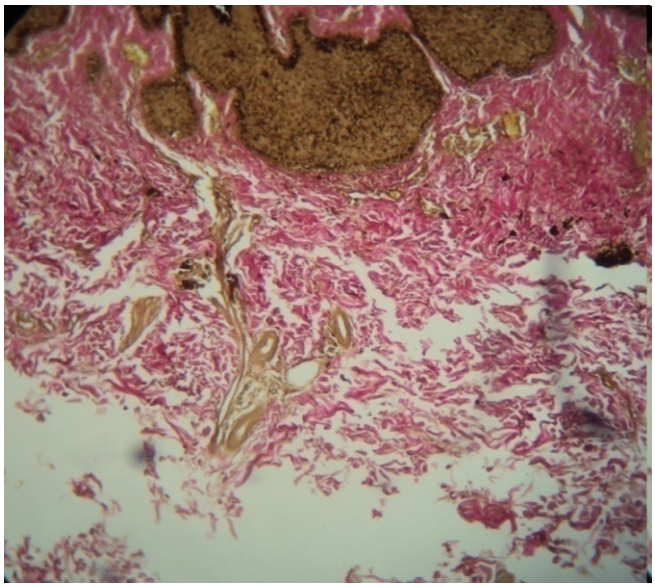

Figure 4:. Increased melanin granules in the basal layer, paucity of elastic fibers in the dermis.(Orcein stain).

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