Journal of Pakistan Association of Dermatologists 2012;**22 (4)**:376-378.

Case Report

Atrophia maculosa varioliformis cutis - a rare case report

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Abstract We present a 4-year-old girl with a 3-year history of spontaneous atrophic lesions of variable shape and size over face. There was no history of trauma, seasonal variation, photosensitivity or presence of inflammatory lesions preceding the development of atrophic lesions over the facial skin. Lesions were increasing in number gradually and spontaneously. Family history was insignificant. On examination sharply defined varioliform lesions i.e. oval, linear and small circular atrophic macules were evident on cheeks, nose and forehead with normal surrounding facial skin. Histopathological examination revealed mild elastic fibre degeneration in dermis. Clinical history, cutaneous and histopathological findings of our case fits in to the description of atrophia maculosa varioliformis cutis.

Key words
Atrophia maculosa varioliformis cutis, spontaneous varioliform scarring of cheek.

Introduction

Atrophia maculosa varioliformis cutis was initially described in 1918 as an entity in which both linear and punctate scars appeared spontaneously on normal facial skin.[1] Atrophia maculosa varioliformis cutis is a rare condition with approximately 20 reports in the literature.[2,3] Typically, spontaneous macular atrophy is reported on the face in the absence of preceding trauma or inflammation but lesions have been described elsewhere on the body. The lesions may be linear, punctate or rounded and some reports do describe initial erythema and scale. The course is usually chronic but slowly progressive. Histological findings are variable and include epidermal thinning, a mild perivascular lymphocytic infiltrate, fragmented elastic fibres and normal or increased collagen. Pathogenesis is unknown and most cases are sporadic, although familial occurrence has been reported.

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Case report

A 4-year-old girl presented with a 3-year history of facial lesions. She developed spontaneous atrophic lesions of variable shape and size over face. There was no history of trauma, varicella or inflammatory lesion preceding the development of atrophic lesion over the facial skin. Lesions were increasing in number gradually and spontaneously. History suggestive of photosensitivity was not present. There was no history of seasonal variation. Family history was not significant. On examination varioliform lesions i.e. oval, linear and small circular atrophic macular scars were evident on cheeks, nose and forehead (**Figures 1, 2**). Macular scars were few mm to 1-2 cm in diameter, shallow and sharply defined. Surrounding facial skin was normal. Histopathological examination revealed normal epidermis and mild elastic fibre

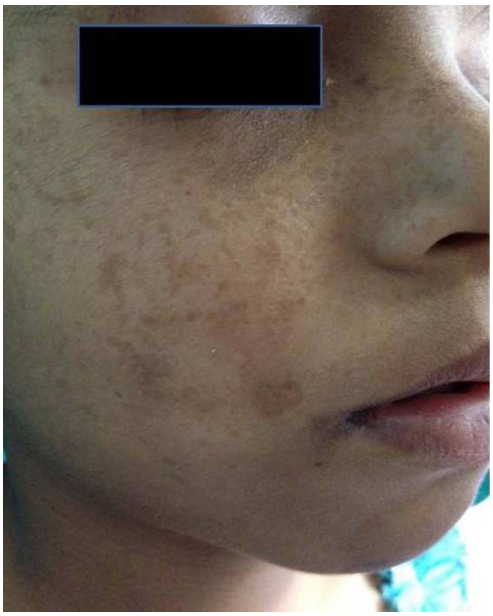

Figure 1 Oval, linear and small circular atrophic macular scars involving right cheek & nose.

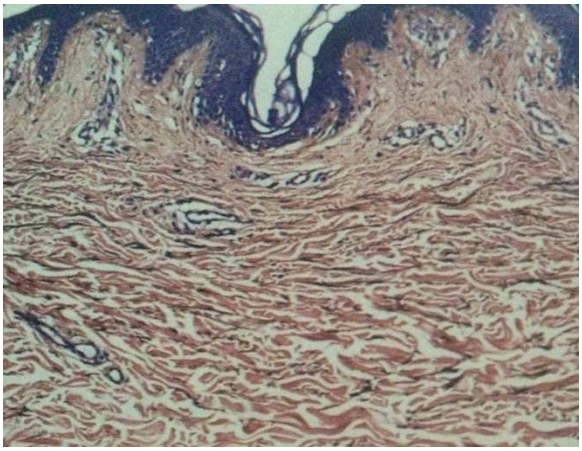

Figure 3 Elastic fibre degeneration in dermis (orcein stain 40x).

degeneration in dermis (**Figure 3**). ANA and screening tests for porphyria were negative.

Discussion

Atrophia maculosa varioliformis cutis was initially described in 1918 as an entity in which both linear and punctate scars appeared spontaneously on normal facial skin. Although its etiology is unknown, atrophia maculosa varioliformis cutis may represent an underlying

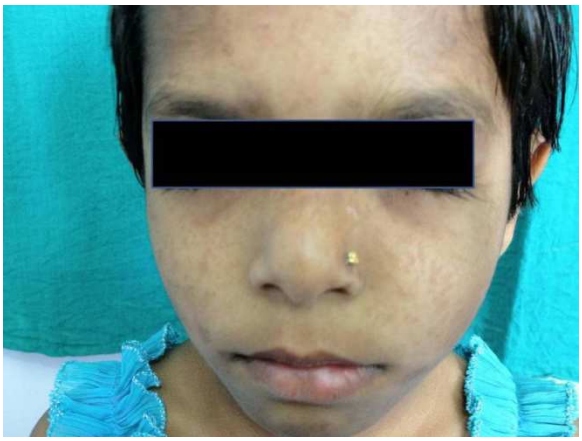

Figure 2 Linear and circular atrophic macular scars involving face.

defect of dermal elastin as demonstrated by histologic and ultrastructural findings.[3] It has been documented only in the skin.

Based on a pedigree assembled by Qu *et al*.[4] atrophia maculosa varioliformis cutis has autosomal dominant inheritance.

Differential diagnoses included scarring secondary to varicella, atrophoderma vermiculatum, hydroa vacciniforme and infantile acne but these were excluded by the chronic course, clinical findings and lack of seasonal variation. Hydroa vacciniforme is a rare idiopathic photodermatosis of childhood characterized by recurrent crops of vesicles over face and other exposed parts, which heals with varioliform scarring. History of photosensitivity and recurrence during summer season will be present in hydroa vacciniforme. Atrophoderma vermiculatum is a rare follicular disorder primarily affecting children with reticular or honeycomb atrophy of the cheeks and forehead. Primary anetoderma is characterized by atrophic macules or patches that may bulge like pouches.

Treatment for atrophia maculosa varioliformis cutis has not been discussed in the literature.

Our case appears to be a sporadic form with spontaneous appearance of varioliform scar without any preceding inflammatory lesion. Lesions have a chronic course with lack of photosensitivity or seasonal variation. Histopathological findings indicate abnormality in elastic fibre.

To the best of our knowledge, only few cases of atrophia maculosa varioliformis cutis have been described in world's literature. We describe a case of 4 year old female child, whose clinical history, cutaneous and histopathological findings fit into the description of atrophia maculosa varioliformis cutis.

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