Atrophia Maculosa Varioliformis Cutis: A Rare Case Report

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Sir,

Atrophia Maculosa Varioliformis Cutis (AMVC) is a non-inflammatory macular atrophic facial dermatosis of young adults. Heidingsfeld was the first to describe this clinical entity in 1918.[1] He used this term to describe the asymptomatic atrophic lesions with clear cut edges on the face of a 20-year-old man. The exact aetiopathogenesis of this disorder is still not clear. Most cases are sporadic. Here we report a case of AMVC, diagnosed based on patient history, clinical feature and histopathological finding.

An 11-year-old boy, presented to us with multiple linear depressed lesions on face since 7 years. On examination, multiple, randomly scattered linear and curvilinear atrophic lesions on bilateral cheeks, angle of the mouth, and nasal bone with length ranging from 1.5 to 3 cm, diameter 0.3 cm and subjective depth 0.1 cm were found [Figure 1]. Lesions were more in number and more prominent on the right side [Figure 2a] than the left side [Figure 2b] of the face. Lesions were of normal skin colour, sharply demarcated and there was no erythema, tenderness, pruritus or scaling. On palpation, lesions were non-compressible and there was no herniation or induration. Family members reported a spontaneous onset of this condition, initially arose at the angles of the mouth, and then increased in number slowly without any symptoms, not preceded by trauma or any inflammatory lesions. The child had no history of preceding varicella, molluscum contagiosum, or herpes infection. His weight had not changed significantly. There were no milia, comedones, or papules on the face. He had no history of any psychiatric illness or any significant medical problems. He was not using any drugs for any other reason. The rest of the skin, hair, nail, mucosa examination was unremarkable. Similar lesions were not present in siblings and family members.

A biopsy was done and histopathology revealed sparse superficial and deep perivascular lymphocytic infiltrate with flattening of epidermal rete pattern. Reticular dermal collagen was thinned at places and arranged parallel to surface epidermis. There was no fibrosis in the dermis [Figures 3 and 4].

The diagnosis of Atrophia maculosa varioliformis cutis was made based on the clinical pictures, histologic findings, and no history of either trauma or previous inflammatory lesions on site. He was advised 0.05% tretinoin cream for topical application and is under follow-up.

AMVC is a rare form of facial non-inflammatory macular atrophic dermatosis. The exact aetiopathogenesis of this condition is still unknown. Cases are mostly sporadic. However, a few familial occurrences have been reported,[2,3] suggesting an inherited disorder. This disease may represent an underlying defect of dermal elastin.[4]

AMVC is clinically characterized by spontaneous onset of asymptomatic atrophic lesions which may be round or oval, linear, curvilinear or rectangular with regular margin and clear cut edges mainly on the face of adolescents. The bilateral malar regions are the most frequently involved site. However, the forehead, mandibular area can also be affected. Typical of this condition is the absence of previous inflammatory or traumatic events at the site of lesion.[3-6] Extrafacial involvement like lesions on periumbilical area, posterior aspect of pinna,[3] and on

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Access this article online

Website: www.idoj.in

DOI: 10.4103/idoj.IDOJ_270_20

Quick Response Code:

How to cite this article: Sethy M, Sachan S, Srinivas CR, Sahu S. Atrophia maculosa varioliformis cutis: A rare case report. Indian Dermatol Online J 2021;12:346-8.

Received: 18-Apr-2020. **Revised:** 19-May-2020. **Accepted:** 22-Jun-2020. **Published:** 28-Sep-2020

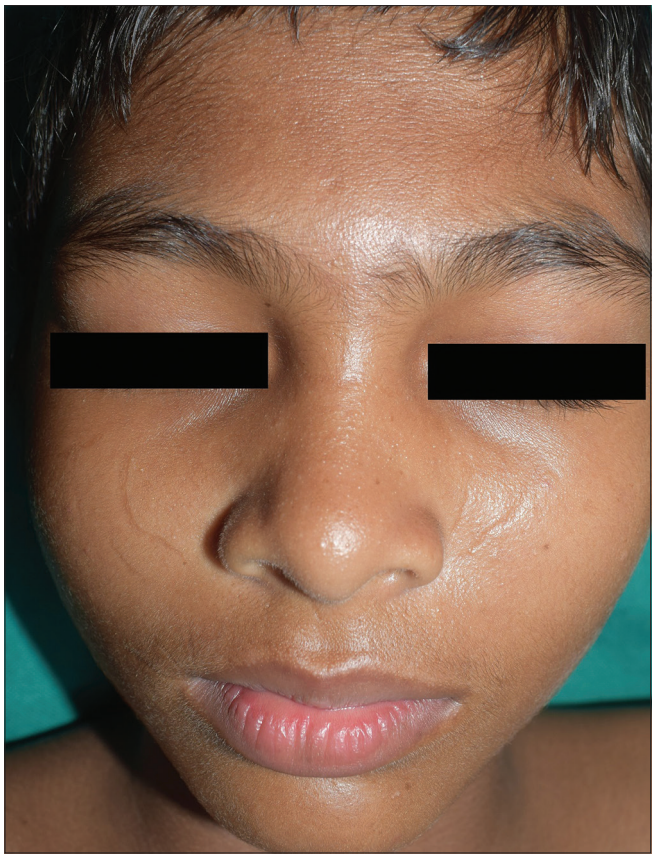

Figure 1: Multiple well-defined linear and curvilinear atrophic lesions with sharp margin on the face

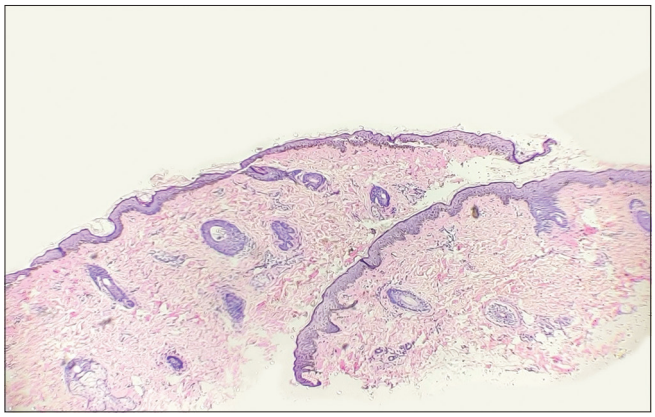

Figure 3: Flattening of epidermal rete pattern, sparse superficial and deep perivascular lymphocytic infiltrate with thinned reticular dermal collagen. (H&E, 4×, 1761 × 1111)

forearm[7] has been reported. The size of the lesion may vary from 0.2-2 cm in length, 0.2-0.5 cm in width; there may not be any pigmentary change or exfoliation of the lesion. Compared to the surrounding skin, they are depressed and cicatricial. Association of AMVC with other disorder is not clear. However, AMVC associated with extrahepatic biliary atresia[8] and pachydermodactyly,[9] and keratosis pilaris[5] has been described previously which may be coincidental. This clinical entity must be

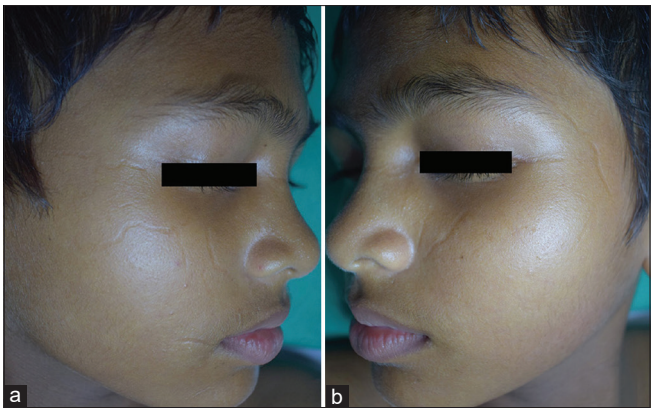

Figure 2: Well-defined linear and curvilinear atrophic lesions with sharp margin on the right side (a) and the left side (b) of the face

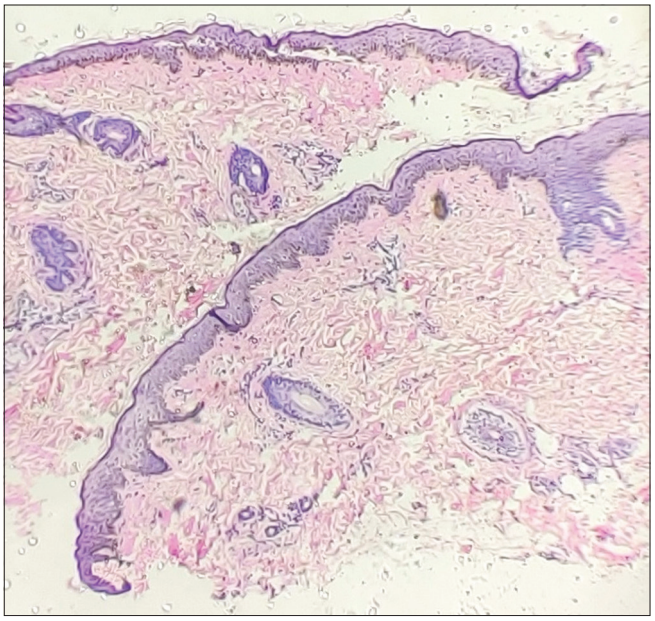

Figure 4: High power view showing flattening of epidermal rete pattern with thinned reticular dermal collagen. (H&E, 40×, 1472 × 1392)

differentiated from scars due to acne, varicella zoster infection, and artifactitious dermatitis.

Histologically common features have been identified include thinning of the horny layer or the entire epidermis, with an apparently normal dermis or a decrease in elastic fibres in the dermis, and a variable lymphocytic perivascular infiltrate in the dermis.[4,5,10]

As no specific and curative therapeutic options are available at present for the management of AMVC, a few drugs like topical and oral retinoids have been tried. Newer treatment modalities like dermabrasion, collagen injections, or laser resurfacing techniques may help in improving facial scarring.

AMVC is an under-reported and mysterious entity. We report this case because of the rarity of this condition and to strengthen and extend the understanding of AMVC.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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