DERMATOPATHOLOGY

REDIFINING ATROPHIA MACULOSA VARIOLIFORMIS CUTIS: A RARE CLINICO-PATHOLOGICAL VARIANT OF SCLERODERMA?

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Background: Atrophia maculosa varioliformis cutis (AMVC) is a rare form of idiopathic non-inflammatory macular atrophy that occurs in young individuals. It is characterized by the spontaneous development of sharply demarcated depressions in a varioliform or linear arrangement. It usually appears on the face, and to date only two cases of extrafacial involvement have been described. A familial occurrence has been reported, suggesting an inherited disorder rather than a response to an environmental insult. The diagnosis is made mainly on a clinical basis. Histology is nonspecific, with a slight decrease of elastic fibers within the papillary dermis, with no or subtle increase of the dermal collagen fibers. Although some authors consider this entity as part of the spectrum of elastic tissue disorders, its pathogenesis remains unknown and its exact nosology is still a matter of conjecture.

Observations: an otherwise healthy 35 years old Moroccan female presented with facial and extrafacial atrophic lesions. The lesions started appearing one year before, and she denied any previous trauma or inflammation. A dermatological examination revealed a bilateral distribution of a few linear, depressed lesions on the cheeks and varioliform, depressed lesions on the trunk and feet. Histology of a biopsy taken from a lesion on a foot showed a morphea-like deposition of abundant sclerotic collagen bundles within the papillary and reticular dermis. Using the Orcein stain, the elastic fibers within the papillary dermis appeared only slightly decreased.

Key message: We report a unique case of AMVC with facial and extrafacial involvement with a peculiar scleroderma-like histological presentation. The dermal sclerosis may be held responsible for the clinically observed epidermal retraction. Our case arises the hypothesis that this rare condition could exists as variant along the spectrum of cutaneous scleroderma.