Dermatology Practical & Conceptual

Necrobiosis-Lipoidica-Like Skin Lesions in a Patient with Non-Systemic Sarcoidosis

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Introduction

Ulcerative-atrophic cutaneous sarcoidosis (CuS) represent a rare form of sarcoidosis, showing different clinicopathological variants: morpheaform, necrobiosis lipoidica (NL)-like and lipodermatosclerosis-like [1]. NL-like CuS is characterized by round, depressed skin lesions with hypopigmentation and telangiectasias, showing histopathological features of both NL and CuS [1]. Only NL has been associated to diabetic microangiopathy [2].

Case Presentation

A non-diabetic-63-year-old Caucasian female came to our attention for a one-year history of painful violaceous erythematous plaques on the left forearm and elbow with ipsilateral elbow joint tenderness. Six months earlier she underwent left elbow MRI and olecranic bone biopsy, revealing a chronic inflammatory process with focal epithelioid non-necrotizing granulomas and negative cultural test. Furthermore, the patient was taking levothyroxine for Hashimoto thyroiditis.

At physical examination the patient presented multiple red-purple atrophic plaques of the left elbow without joint tenderness (Figure 1A). Upon dermoscopy, the lesions showed a heterogeneous orange-pinkish background with white structureless areas and follicular keratotic plugs. The lesions vascular pattern was characterized by linear irregular, arborizing vessels (Figure 1 B and C) and peripheral hairpin vessels (Figure 1C).

Skin punch-biopsy revealed numerous non-necrotizing epithelioid granulomas and Langhans giant cells, located in the superficial and deep dermis. Granulomas were

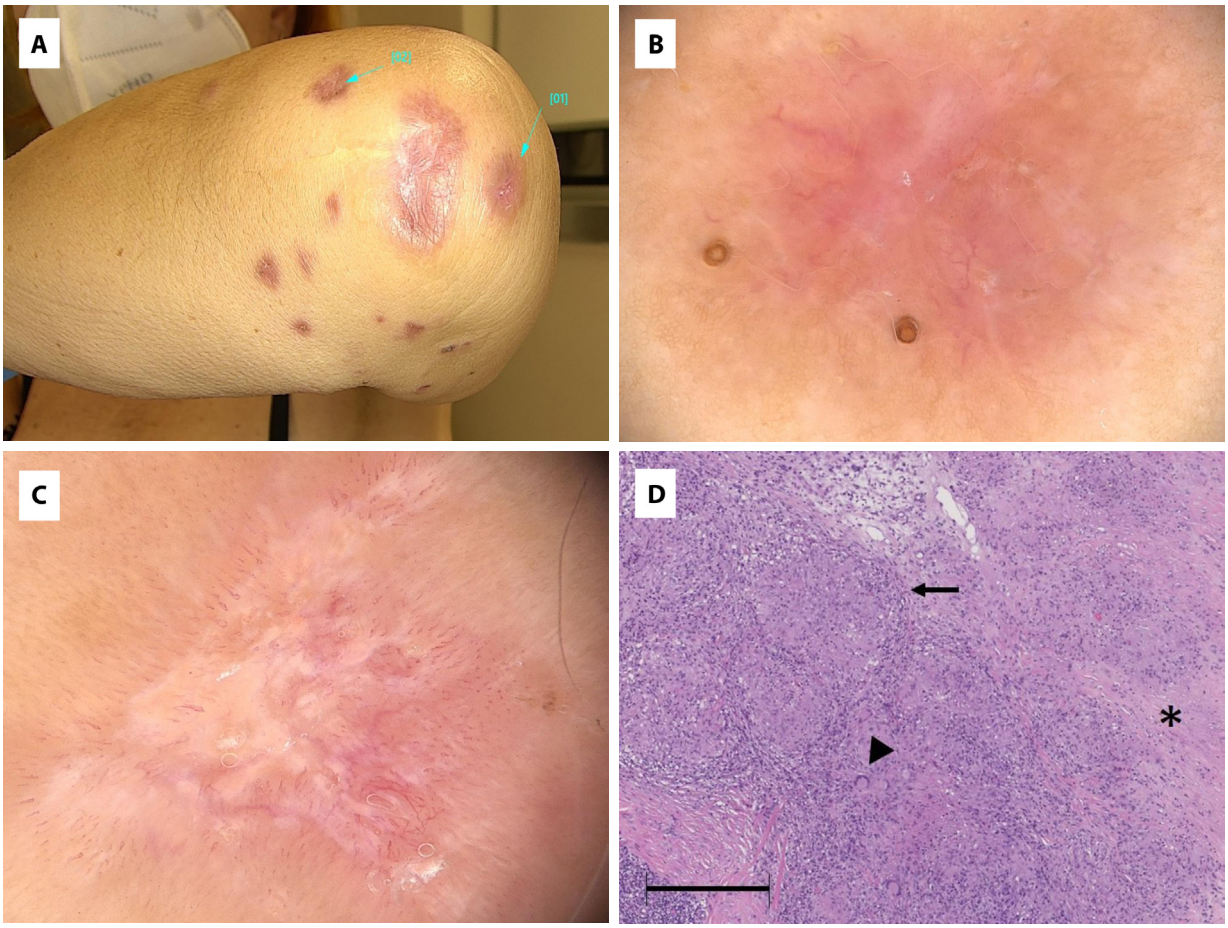

Figure 1. (A) multiple red-purple ulcerated plaques of the left elbow, associated with joint pain. (B,C) Dermoscopy showed a heterogeneous orange, pinkish, reddish background with white structureless areas and aspecific vascular pattern: linear irregular, arborizing vessels plus peripheral haipin vessels. (D) Tissue slide showing non-necrotizing epithelioid granulomas (arrow) with Langhans giant cells (arrowhead) surrounded by lymphocytes. Note the areas of necrobiosis (asterisk) (H&E stain, 5x, scale bar: 500 µm).

surrounded by intense lymphocytic infiltrate with numerous plasma cells. Moreover, palisading histocytes around areas of necrobiosis were also prominent (Figure 1D). Other causative agents for granuloma formation (ie foreign body, microorganisms) were clinically excluded and supported by negative Periodic Acid-Schiff (PAS) and Ziehl-Neelsen special stains. In conclusion the clinical, dermoscopic and histopathological findings plus the negative systemic assessment (ie thoracic-CT, pulmonary function test, electrocardiogram, serum ACE, CRP, VES), supported the diagnosis of localized NL-like CuS with previous ipsilateral bone sarcoidosis. Topical betamethasone cream applied twice daily was prescribed. At four-months follow up the lesions appeared smaller, less infiltrated with reduction of referred pain and negative blood tests.

Conclusions

Ulcerative/atrophic CuS and NL are granulomatous-skin disorders of unknown etiology, sharing similar clinicopathological features [2,3]. As for our case, NL-like CuS affects middle-aged females, without history of diabetes/glucose intolerance [2,3]. In our patient, the lesions were located at the upper limbs, while NL and NL-like CuS affect mostly the lower limbs [3]. The coexistence of CuS with NL in the same lesion underlie the possibility that these represent different stages of the same granulomatous skin disease [3].[3]

Dermoscopy is useful for differential diagnosis of granulomatous skin diseases [4]. NL presents heterogeneous orange-pinkish background and comma/serpentine/arborizing vessels [5]. On the other hand, CuS presents a pink homogeneous background, white structureless areas and fine white scales with disease stage-dependent vascular patterns [4,5]. Our NL-like CuS case showed mixed dermoscopic findings: background color compatible with NL, white structureless areas typical of CuS, and arborizing plus peripheral hairpin vessels compatible with both CuS and NL [4,5].

To the best of our knowledge, this is the first case of upper limb NL-like CuS associated to localized sarcoidosis. This CuS variant is associated to systemic sarcoidosis; thus, complete staging and strict follow up is crucial for treatment.[2]

First line treatment for localized NL-like CuS is based on topical/intralesional corticosteroids, while oral prednisone should be considered for recalcitrant/systemic disease [2,4].

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