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Recommended Citation

Mutizwa, Misha M.; Tang, Mark B.; and Ng, See Ket, "Prominent Degos-like skin lesions in a patient with chronic cutaneous lupus erythematosus." *Dermatology Online Journal*.16,7.. (2010).
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Prominent Degos-like skin lesions in a patient with chronic cutaneous lupus erythematosus

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Dermatology Online Journal 16 (7): 5

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Abstract

Malignant atrophic papulosis, commonly known as Degos disease, is a rare vasculopathy encompassing both benign, cutaneous and lethal systemic variants. We report a case of chronic cutaneous lupus erythematosus in a 41-year-old male presenting with prominent Degos-like skin lesions. Multiple atrophic, porcelain-white, scar-like papules and plaques with dusky, erythematous borders, suggestive of malignant atrophic papulosis, were noted on the patient's back. Additional cutaneous findings included photo-distributed facial erythema and discoid lupus-like plaques on the face, shoulders, and arms. Clinicopathological correlation supported a diagnosis of chronic cutaneous lupus erythematosus; hydroxychloroquine was initiated with good clinical response. No new or active lesions were observed at the sixteen-month follow-up. This case highlights a rare skin finding associated with chronic cutaneous lupus erythematosus and underscores the importance of ruling out primary autoimmune disease, particularly lupus, before a diagnosis of malignant atrophic papulosis can be made.

Introduction

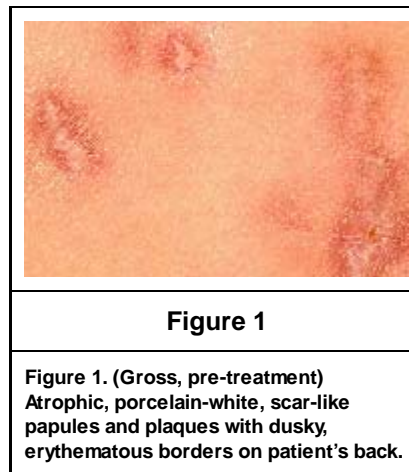
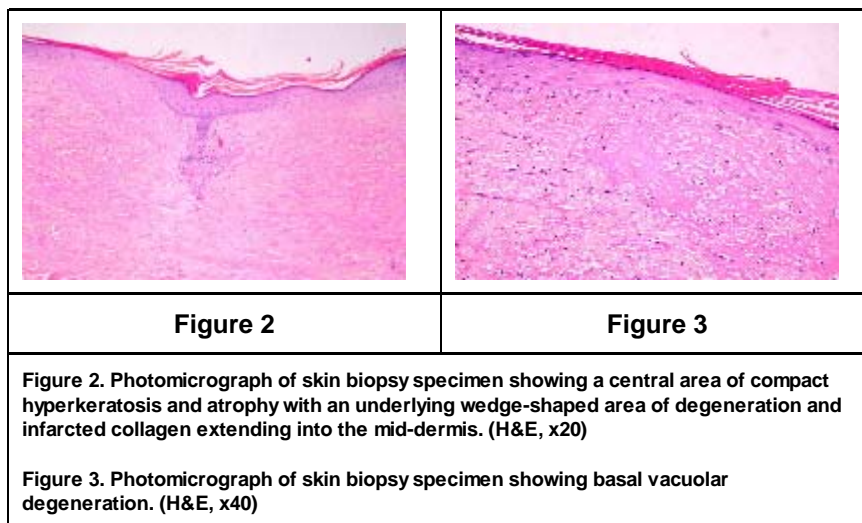
Malignant atrophic papulosis (MAP), commonly known as Degos disease, is a rare, occlusive microangiopathic vasculopathy characterized by the presence of "pathognomonic" skin lesions, commonly described as scarred papules with atrophic, porcelain-white centers and a rim of peripheral erythema and telangiectasia [1]. Classically-defined MAP is multisystemic and almost universally fatal [1]. Whereas a variety of organ systems can become involved, gastrointestinal and neurologic sequelae are most common. Death most often results from bowel perforation [1]. A strictly cutaneous variant of MAP has also been recognized and portends a more favorable prognosis [1].

Degos-like skin lesions have, on occasion, been reported in association with autoimmune disorders such as lupus erythematosus, progressive systemic sclerosis, Wegener granulomatosis, and dermatomyositis [2, 3, 4, 5]. We report a rare case of chronic cutaneous lupus erythematosus (CCLE) associated with prominent Degos-like skin lesions, highlighting the possibility that these uncommon skin lesions may be an atypical presenting sign of autoimmune skin diseases such as CCLE.

Case report

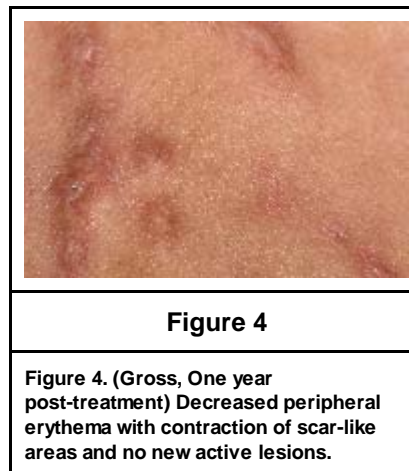
A 41-year-old male with no notable past medical history presented with widespread papular skin lesions of three years duration on his back. Cutaneous exam revealed prominent atrophic, porcelain-white, scar-like papules and plaques with dusky, erythematous borders and areas of confluence on the back (Figure 1). Additional cutaneous findings included photo-distributed facial erythema and scattered erythematous, scaly papules and plaques over

Baseline laboratory investigation was notable for a positive anti-nuclear antibody titer of 1:400 with a speckled, homogenous pattern. Additional autoimmune markers were negative, including autoantibodies against double-stranded DNA, extractable nuclear antigens, cardiolipin, and phospholipids.



Histopathological examination of a lesion from the patient's back showed a central area of compact hyperkeratosis and atrophy with an underlying wedge-shaped area of degeneration and infarcted collagen extending into the mid-dermis (Figure 2), consistent with MAP. Additional histological findings included vacuolar basal degeneration (Figure 3), a dermal perivascular and perifollicular lymphocytic infiltrate, and increased mucin deposition identified with alcian blue staining. Direct immunofluorescence studies were negative.

A presumptive diagnosis of CCLE was made after careful clinicopathological correlation. The patient was started on hydroxychloroquine (5 mg/kg/day) and low-dose aspirin. Smoking cessation was also advised. On follow-up, there was decreased peripheral erythema and contraction of the scar-like areas in the lesions on the patient's back (Figure 4). The erythematous papules and plaques improved without scarring. At a sixteen-month follow-up, the patient remained systemically well with no new or active cutaneous lesions having developed since the initiation of therapy.



Discussion

Whereas the cutaneous findings in lupus are myriad, this report illustrates an exceedingly rare case of Degos-like lesions as the first presenting sign of CCLE. The importance of distinguishing between cutaneous lupus and MAP cannot be overemphasized because MAP is associated with systemic involvement and poor response to treatment. Death most commonly occurs within two to three years of diagnosis [1].

This patient's diagnosis of CCLE was based on careful clinicopathological correlation. Whereas his back lesions were the presenting complaint, a complete cutaneous examination revealed the additional findings of photodistributed erythema and discoid lupus-like plaques on the face, shoulders, arms, and thighs. Other evidence supporting the diagnosis was the patient's positive ANA titer and pattern and his prompt, favorable response to hydroxychloroquine, a mainstay of CCLE therapy. Because unresponsiveness to therapy has been described as one of the defining characteristics of MAP, the fact that the patient's Degos-like lesions and lupus-like plaques simultaneously improved upon initiation of hydroxychloroquine is also supportive of the diagnosis of CCLE [6].

Malignant atrophic papulosis is an occlusive, small-vessel vasculopathy that results in tissue infarction [1]. Accordingly, histological examination in patients with MAP commonly reveals wedge-shaped areas of degenerated, infarcted collagen, as seen in a biopsy from this patient's back [1]. Whereas this form of wedge-shaped necrosis is not typically seen in CCLE, the additional findings of basal vacuolation, an interface dermatitis, a perivascular lymphocytic infiltration, and increased mucin deposition lent support to its diagnosis. These latter histological findings are characteristic of lupus, but they can also be observed in MAP, somewhat limiting the usefulness of histopathology in distinguishing between these clinical entities [7]. Nonetheless, histopathology is important in ruling out other possibilities, such as atrophie blanche, which has also been described in the setting of lupus and can clinically resemble the lesions characteristic of MAP [8].

It is interesting to note that this patient's Degos-like lesions showed areas of confluence. This is an unusual finding that is not described in classical MAP, in which the representative lesions are typically discrete, well-demarcated, atrophic papules. We believe that this distinct clinical feature of lesional confluence may be useful in differentiating between classical MAP and other causes of Degos-like lesions. Indeed, a similar example of coalescing Degos-like lesions has been observed in a patient with underlying dermatomyositis [7].

Given the likelihood of an underlying occlusive microangiopathic process, the patient was treated with a combination of hydroxychloroquine and low-dose aspirin. Anti-platelet therapy has been reported to be useful in the benign cutaneous variant of MAP, although clinical response is highly variable [9, 10]. In addition to its immunomodulatory effects, hydroxychloroquine has been found to have important antithrombotic properties, making it a potentially ideal therapeutic agent for such cases of CCLE. Hydroxychloroquine has been postulated to derive its antithrombotic effects through multiple mechanisms, including the inhibition of platelet aggregation and adhesion and the lowering of cholesterol levels [11]. It has also been shown to interfere with antiphospholipid (aPL) antibody production, suggesting that the drug may be especially appropriate in cases of aPL positive lupus [12]. A recent retrospective study analyzing risk factors for thrombosis in a large, multi-ethnic systemic lupus erythematosus cohort was the first clinical study to confirm hydroxychloroquine's protective anti-thrombotic effects [13]. Our patient was also advised to stop smoking, given the positive association between smoking and thrombosis and evidence that smoking limits the effectiveness of antimalarial therapy in patients with CCLE [14].

It is interesting to note that whereas the association between Degos-like lesions and lupus has previously been described, these reports typically describe patients with systemic lupus erythematosus (SLE) rather than CCLE. Whereas some of these patients with SLE are likely to have had discoid lesions, to our knowledge, this is the first report of Degos-like lesions as the presenting complaint in a patient with characteristic discoid lesions who did not meet the criteria for a diagnosis of SLE. Although it is possible that this patient will, in the future, develop additional clinical or laboratory findings that would qualify for him a diagnosis of SLE, the ultimate classification of this patient within the family of lupus diseases is of secondary importance. What is more significant about this case is the fact that it underscores the importance of excluding treatable conditions associated with Degos-like lesions given the marked differences in therapy and prognosis for patients. This case demonstrates that primary autoimmune disease, and in particular, lupus, must be excluded before a diagnosis of MAP can be made.

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