Subcutaneous Sarcoidosis Resembling Cellulitis

To the Editor:

Sarcoidosis is a systemic disease, and skin manifestations develop in 25% of cases.1 The skin lesions, which vary greatly, are classified as either specific lesions, which show noncaseating granulomas on skin biopsy, or nonspecific lesions, which show no histological evidence of these granulomas.1 Subcutaneous lesions account for 1.4% to 6% of specific lesions, depending on the series.2,3

Although skin involvement is thought not to be prognostically significant or to correlate with the presence of systemic disease,1 a strong association has been observed between the appearance of subcutaneous lesions and the development of systemic disease.2

We describe the case of an 81-year-old woman with no past history of relevance, in whose left forearm a painful erythematous indurated plaque had developed after a sting. The initial diagnosis was bacterial cellulitis (Figure 1A); however, when it was observed that the lesion failed to respond to antibiotic and anti-inflammatory treatment, biopsy was performed that included subcutaneous tissue. Histology showed the epidermis, superficial dermis, and mid dermis to be intact, but revealed, in the deep dermis and in the adipose tissue (Figure 2A), an intense inflammatory reaction consisting of giant cells with central fibrinoid tissue and peripheral lymphocytes (Figure 2B). Polarized light did not reveal any foreign material, and Giemsa, Ziehl-Neelsen, and methenamine silver stains were all negative. Mycobacterial culture on Lowenstein-Jensen medium was also negative.

Additional tests revealed enlarged mediastinal lymph nodes compressing the lower third of the esophagus. Laboratory tests showed angiotensin-converting enzyme (ACE) levels of 92 U/L and an erythrocyte sedimentation rate (ESR) of 50 mm/h. The Mantoux test was negative. Since all findings were consistent with a diagnosis of stage I systemic sarcoidosis, the treatment prescribed was oral prednisone, 45 mg once daily, and hydroxychloroquine, 100 mg twice daily. After 6 months, the lymphadenopathies had significantly reduced in size and the skin lesions had cleared (Figure 1B); after 12 months, the corticosteroid and antimalarial treatment was withdrawn. After 2 years of follow-up, no new skin lesions had appeared and the mediastinal lymph nodes measured less than 1 cm.

Subcutaneous sarcoidosis predominantly affects women, and peak incidence occurs in the fourth decade of life.4 The literature reflects a close association between subcutaneous sarcoidosis and systemic symptoms, and so it is unusual to see exclusively subcutaneous involvement. The subcutaneous symptoms typically present in the form of nodules3 or plaques. A presentation that masquerades as cellulitis is rare and can consequently delay diagnosis—as happened in our case.4 This is because the fusion or merging of multiple firm and elastic subcutaneous nodules arranged in linear bands can be confused with the kind of indurated plaque that is suggestive of cellulitis.5

Once a bacterial, fungal, or viral cause is ruled out, diagnosis of subcutaneous sarcoidosis is based on Vainsencher and Winkelmann criteria,6 namely, a histologic finding of sarcoïdal granulomas with minimal lymphocytic infiltrate in the adipose tissue.

Oral corticosteroids are the treatment of choice for subcutaneous sarcoidosis as they result in an excellent response, whereas the response to second-line drugs such as antimalarials tends to be incomplete. The combined treatment with corticosteroids and antimalarial drugs that we opted for from the outset resulted in an early and favorable response in our patient. In a study of 21 patients with subcutaneous sarcoidosis, treatment with corticosteroids at daily doses of between 20 mg and 40 mg led to a response in 80% of patients within the first 4 to 8 weeks.4

Subcutaneous sarcoidosis is a subtype with specific characteristics, one of which is presentation with nodules or plaques that mimic cellulitis. Although subcutaneous sarcoidosis responds well to treatment with corticosteroids, this variant of sarcoidosis correlates closely with systemic involvement, with the resulting implications for disease prognosis.
Figure 1  Subcutaneous sarcoidosis plaque located on the left forearm. A, before treatment. B, after 3 months of treatment with oral corticosteroids and antimalarial drugs.

Figure 2  A, inflammatory reaction in the deep dermis and in the subcutaneous cell tissue (hematoxylin-eosin, original magnification ×20). B, higher magnification of the sarcoidal granulomas (periodic acid-Schiff, original magnification ×100).

References


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