Annular Erythema Associated with Anti-Ro/SS-A and Anti-La/SS-B Antibodies: a New Case in a Hispanic Patient

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We describe an 18 year-old male patient who was anti-Ro/SS-A and anti-La/SS-B positive and presented with recurrent annular plaques on the trunk, arms, face, and scalp with evidence of associated patchy alopecia. The skin biopsy revealed a superficial perivascular and periappendegeal lymphocytic infiltrate and focal areas of vacuolar alteration and smudging of the dermoepidermal junction. The patient also presented with a history of xerophthalmia. Skin lesions as well as sicca symptoms responded to antimalarial treatment with hydroxychloroquine. This case demonstrates a new case of annular erythema indistinguishable from those previously described in patients of Asian descent occurring in a Hispanic patient.

Key words: Annular erythema, Sjögrens syndrome, Lupus erythematosus

Annular erythemas may be associated with a several immunologic abnormalities including subacute cutaneous lupus erythematosus (1), autoimmune annular erythema (2), and neonatal lupus erythematosus (3). More recently, a unique, recurrent annular erythema in anti-Ro/SS-A and/or anti-La/SS-B positive patients with clinical or subclinical features of Sjögren's syndrome (SS) has been reported in patients of Asian descent (4-5). These lesions have also been described in patients with positive anti-Ro/SS-A and/or anti-La/SS-B that fulfill the diagnostic criteria for systemic lupus erythematosus (SLE) (6-7).

Only two cases in patients of non-Asian descent have been reported in the English literature (8-9). We report the case of an 18-year-old Puerto Rican male with positive anti-Ro/SS-A and anti-La/SS-B antibodies, lesions of annular erythema and symptoms of Sjögren's syndrome.

Case report

An 18 year-old male patient with no history of systemic illness was seen in our clinics on 2006 with a complaint of asymptomatic annular lesions present on the face and upper extremities of several months duration. Upon physical examination, the patient was found to have multiple annular, erythematous plaques with raised borders and no scaling over bilateral arms and one larger plaque over the forehead. A skin biopsy was performed with a presumptive diagnosis of granuloma annulare. The biopsy revealed a mild superficial perivascular lymphohistiocytic infiltrate, scattered melanophages in the papillary dermis and focal areas with smudging of the dermoepidermal junction. Alcian blue stain for mucin was negative. Topical corticosteroids had partial improvement of skin lesions, and patient referred that some lesions disappeared without scarring, while new lesions eventually appeared.

Two years later the patient presented with extensive lesions on the trunk, bilateral arms, face, and scalp. (Figure 1) Annular plaques present on the scalp resulted in patchy alopetic patches with no scaling or evidence of atrophy. The skin lesions were erythematous, annular plaques with raised borders and central clearing. At this moment, due to the presence of scalp lesions with associated alopecia a skin biopsy was repeated. A biopsy revealed a superficial perivascular and periappendegeal lymphocytic infiltrate associated with focal areas of vacuolar alteration and blunting of the dermoepidermal junction. (Figure 2) The scalp biopsy revelaed a scant superficial perivascular and periappendegeal lymphocytic infiltrate, also with focal areas of vacuolar alteration of the dermoepidermal junction and melanophages in the papillary dermis. (Figure 3) Direct immunoflourescence studies for IgG, IgM IgA, and C3 were negative in both the scalp and skin biopsy specimens.

Laboratories revealed a positive antinuclear antiboby (1:640) with a speckled and nucleolar pattern. Anti-Ro/SS-A and anti-La/SS-B were also positive with titers >100 EU/ml. Other laboratories including complete blood

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The authors have no conflict of interest to disclose.

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count, comprehensive metabolic panel, anti-dsDNA, rheumatoid factor, and C3 and C4 levels were within normal limits.

Upon further questioning, the patient admitted having symptoms of xerophthalmia since he was 12 years old which were being treated with lubricant eye drops. He denied symptoms of xerostomia. After ophthalmologist and rheumatologist evaluation, patient was started on oral hydroxychloroquine alternating every other day between 200mg and 400mg without complications. After 2 months of treatment all his skin and scalp lesions had resolved and he was exhibiting regrowth of hair over alopetic patches.

Discussion

A variety of skin lesions may be associated with Sjogren's syndrome, including hypergammaglobulinemic purpura, urticarial vasculitis, and autoimmune anhidrosis (5). In the past decades, a rare, distinct annular erythema has been described, mostly in patients of Asian descent with positive anti-Ro/SS-A and/or anti-La/SS-B antibodies and SS or SS/SLE overlap (4-7). It has been called annular erythema of Sjögren's syndrome by some, since patients present with clinical or subclinical symptoms of this condition. We report a case with clinical and histological features indistinguishable from those previously reported of annular erythema associated with anti-Ro and/or anti-La antibodies in a Hispanic patient.

In patients of Asian descent, annular erythema associated with anti-Ro and/or anti-La antibodies is characterized by erythematous, annular, sometimes polycyclic plaques with an elevated edematous border and central pallor (4-5). The lesions occur predominantly on the face (especially the cheek and preauricular area), upper extremities and back (5), as seen in our patient, but cases with widespread involvement have been reported (5, 9). Also, lesions have been reported to occur on the scalp resulting in patchy alopecia (7) similar to our case. Lesions may resolve spontaneously, without scarring, but are usually recurrent (4, 10-11).

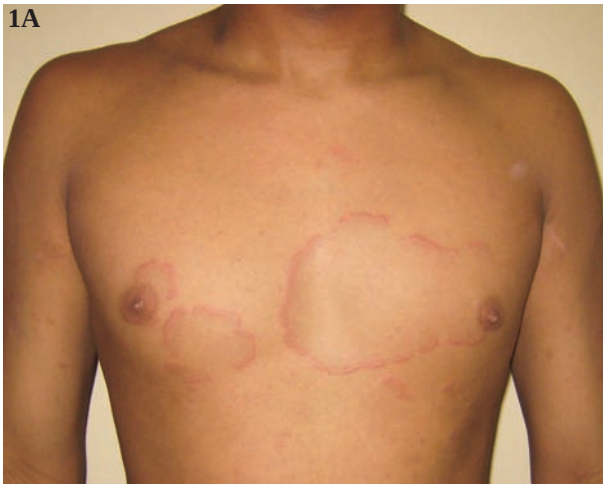
Histologic findings in annular erythema associated with anti-Ro and/or anti-La antibodies include the presence of a perivascular and/or periappendegeal infiltrate of lymphocytes throughout the dermis which may be associated with edema of the papillary dermis (4-5, 7, 10, 12). The epidermis may be normal or may show focal vacuolar degenartion (4-5, 10, 12). Still, other features of lupus erythematosus, such as thickening of the basement membrane, follicular plugging, and epidermal atrophy are not observed (12). In some cases, deposition of immunoglobulin or complement along the dermoepidermal junction has been reported (5, 8, 10, 12).

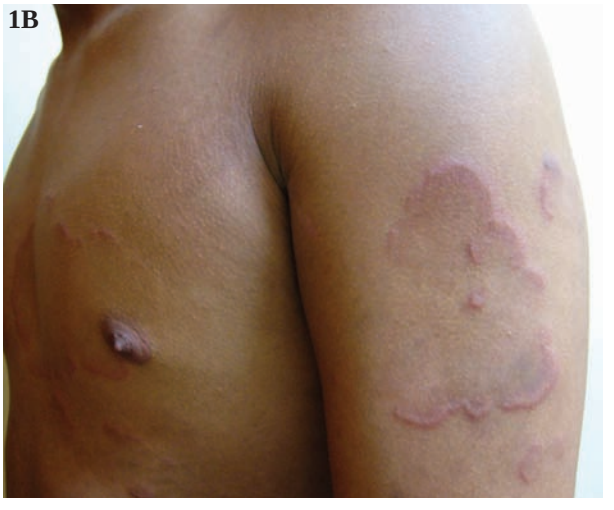
Some authors argue that annular erythema associated with anti-Ro and/or anti-La antibodies represents a distinct clinical entity (4-6) while others suggest it represents a variant of subacute cutaneous lupus erythematosus (7, 10). Nishikawa, et al. (13), indicate that Asian patients with anti-Ro/SS-A antibodies frequently demonstrate plaque-like, indurated, erythematous, doughnut-like lesions clinically distinguishable from the papulosquamous, photosentitive lesions more commonly associated with anti-Ro/SS-A positive Caucasian patients with subacute cutaneous lupus erythematosus (SCLE). They suggest that the antibody response in white patients is associated with a statistically significant increased frequency of HLA-DR2 and/or HLA-DR3 phenotypes. On the other hand, Miyagawa, et al. (14) and Niizeki, et al. (15) have reported a 100% frequency of HLA-DRw52 in Japanese patients with annular erythema. Based on these findings one could speculate that the variability of cutaneous lesions encountered in patients with positive anti-Ro and/or anti-La antibodies may be due to immunogenetic differences in HLA expressions. Some have suggested that annular erythema in SS or SS/SLE among Orientals may be the counterpart of cutaneous lesions of SCLE among Caucasians (10-11).

The clinical and histologic features, laboratory findings, as well as symptoms of Sjögren's syndrome seen in our patient are compatible with the previously reported cases of annular erythema associated with anti-Ro and/or anti-La antibodies. This is, to our knowledge, the first report in a Hispanic patient. Still, more studies are warranted to further characterize this entity and better understand the racial influence in the clinical expression of anti-Ro and/or anti-La positive individuals.

Resumen

Se describe el caso de un paciente de 18 años de edad, con serología positiva para los anticuerpos anti-Ro/SS-A y anti-La/SS-B que presenta con placas anulares recurrentes en el tronco, los brazos, la cara y el cuero cabelludo, con evidencia de alopecia asociada. La biopsia de la piel reveló un infiltrado superficial perivascular y periadnexal linfocítico, y áreas focales de alteración vacuolar y borramiento de la unión dermo-epidermal. El paciente también presentó un historial de xeroftalmia. Las lesiones en la piel, así como los síntomas de sicca, respondieron a tratamiento antimalarial con hidroxicloroquina. Este es un caso novel de eritema anular asociado a los anticuerpos anti-Ro y/o anti-La en un paciente hispano, indistinguible de los anteriormente descritos en los pacientes de origen asiático.





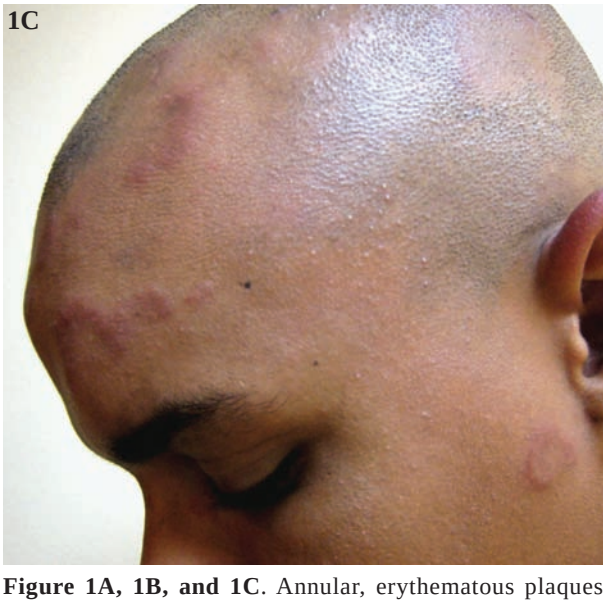
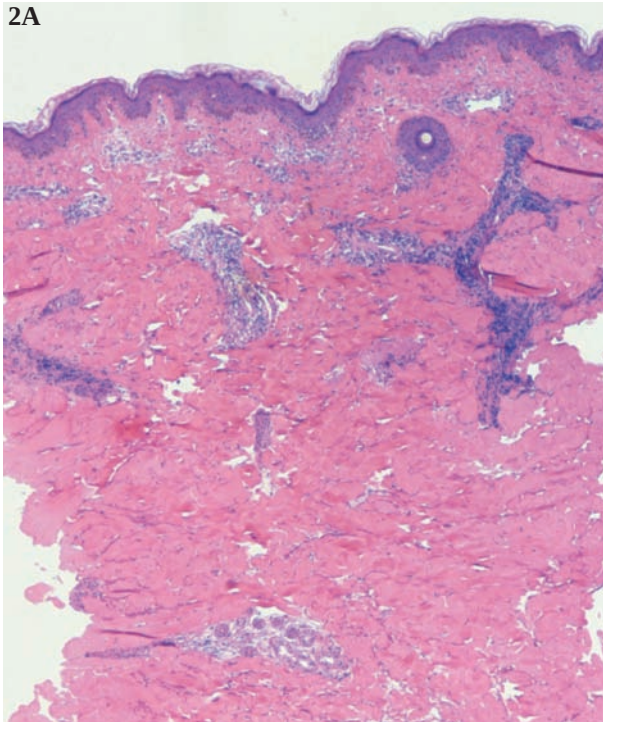


Figure 1A, 1B, and 1C. Annular, erythematous plaques with central clearing on the chest, arms, face, and scalp with associated alopecia.



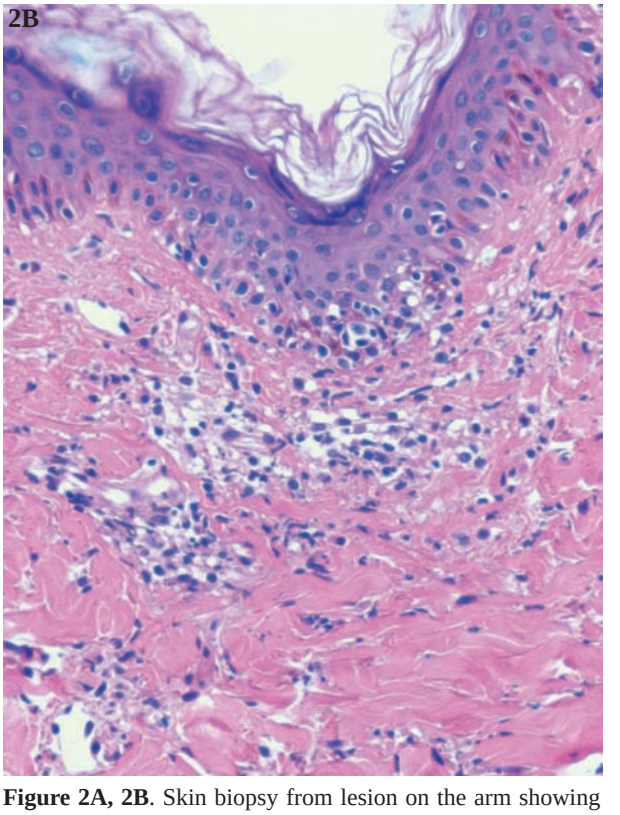


Figure 2A, 2B. Skin biopsy from lesion on the arm showing a perivascular and periappendegeal lymphocytic infiltrate in the superficial and mid-dermis with areas of focal vacuolar alteration in the basal cell layer. (Hematoxylin-eosin stain; original magnification: A, X10; B, X40.)

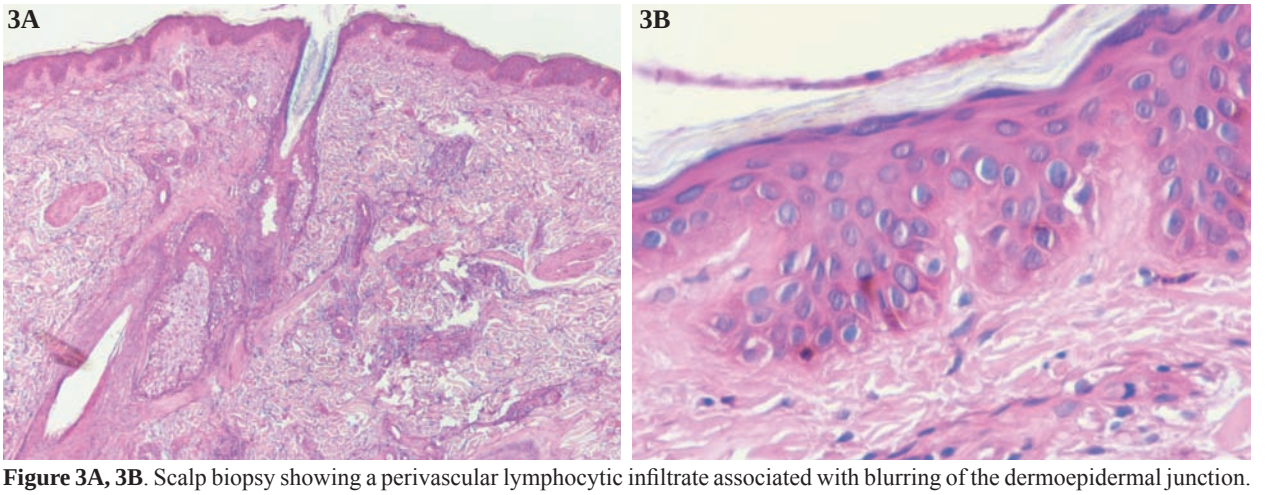


Figure 3A, 3B. Scalp biopsy showing a perivascular lymphocytic infiltrate associated with blurring of the dermoepidermal junction. (Hematoxylin-eosin satin; original magnification: A, X10; B X20.)

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