Persistent Dermal Plaque Lesions in Adult Onset Still's Disease

S.		Kaur ^a
P.		Bambery ^b
S.		Dhar ^a

Departments of ^aDermatology, Venereology and Leprology; ^bInternal Medicine, Post Graduate Institute of Medical Education and Research, Chandigarh, India

Prof. Surrinder Kaur Department of Dermatology. Venereology and Leprology Pgimer, Chandigarh 16(1 012 (India)

The adult onset of Still's disease (AOSD) was accepted as a distinct clinical entity recently [1]. It is defined in clinical terms and the diagnosis is based on meeting certain clinical criteria [2-4], as there is no diagnostic test. Considered rare, it is probably an under-reported cause of fevers of unknown origin [5-7]. As in its paediatric counterpart, patients with AOSD often present with a blotchy, salmon-coloured, maculopapular rash on the trunk and arms. It closely parallels the rise and fall of temperature [5, 7, 8]. This rash is an important diagnostic criterion [2-4]. We recently observed another type of skin lesion in a patient with AOSD. We believe this has not been described previously.

Case Report

A 19-year-old female was symptomatic for the preceeding year with episodes of high fever ($> 39^{\circ}\text{C}$), polyarthralgia and a skin rash. There was no systemic infection. Joint pain initially involved the small joints of the hand but later spread to the large joints symmetrically. There were no signs of inflammation. She complained of two distinct types of skin rash. One was evanescent, pinkish and maculopapular over the trunk and upper arms. It came on with the rise of

temperature and subsided during defervescence. The other was persistent and consisted of erythematous confluent papules that coalesced to form large, irregular, erythematous plaques varying from 3 to 10 cm in diameter. They involved the anterior chest, hypochondriac regions, neck, back, dorsa of the forearms and the shins bilaterally and symmetrically (fig.1). They were mildly pruritic, and, while she was under observation, they spread to her face and increased in size progressively

(fig. 2). A skin biopsy revealed hyperkeratosis, mild acanthosis with spongiosis, mild upper dermal oedema with perivascular and periappendageal lymphomononuclear infiltrate.

She also had pallor, generalised lymphadenopathy, hepatic and splenic enlargement but no serositis, cutaneous vasculitis, throat, eye or neurologic abnormality. Several investigations carried out to identify causes for such a syndrome failed to show any abnormality. Lymph node histology revealed reactive hyperplasia, and the liver biopsy was normal. She received aspirin (4.5 g/day) without relief. The addition of indomethacin,

Fig. 1. Distribution of persistent plaque lesions at the beginning.

naproxen and piroxicam also did not help. Finally she received prednisolone (0.6 mg/kg/ day) and responded very well. All manifestations disappeared within 3 weeks. Corticosteroids were gradually tapered off over 6 months. When last seen, she had been well for more than a year off all medication.

Discussion

Surprisingly, Still did not document the characteristic rash now recognised as an important diagnostic criterion for the condition that bears his name [9]. It was described more than 50 years later [8]. Our patient had the typical rash but in addition had another

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which has not been documented. She met the diagnostic criteria for AOSD otherwise [2-4]. The second dermal lesion had a non-specific histology, worsened with the systemic disease and responded to treatment. This evolution suggested that it may be a manifestation of the primary process. Its recurrence during relapse would further strengthen this argument. Other dermatologic conditions which may mimic this rash include the lichenified plaque of atopic dermatitis, erythroderma variabilis and lichen simplex chronicus. However, she did not have evidence of any of these and had not used any topical therapy for her joint pain or rash.

AOSD has two distinct clinical courses [10]. One is monophasic, usually very responsive to aspirin or a NSAID and not complicated by any serious organ dysfunction. The other is polyphasic, with frequent relapses, requires corticosteroid therapy and is often complicated by amyloidosis, visceral and articular disease [7]. So far, our patient appears to have the former. Fig. 2. Distribution of the same lesions during the period of observation.

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