Clinical and Experimental Dermatology

Erythematous plaques on the face: leonine facies

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doi:10.1111/j.1365-2230.2008.02820.x

Clinical findings

A 20-year-old woman presented with erythema and swelling on her face, which had progressively increased during the previous 4 months. There were erythematous, infiltrated, tiny scaled plaques on the forehead, nasal area, cheeks, chin and perioral area, which gave the patient a leonine appearance (Fig. 1).

There was no lymphadenopathy or hepatomegaly. Results of radiography of the chest and hands, electrocardiography, echocardiography, ocular examination, complete blood cell count, biochemical tests, syphilis serology tests and tuberculin skin test were negative or within normal limits. Tests for serum angiotensin-converting enzyme cannot be carried out in our hospital.

Histopathological findings

A skin biopsy was taken, which showed multiple, small, well-circumscribed noncaseating epithelioid granulomas with Langerhans' giant cells, in association with a few lymphocytes (Fig. 2). Ziehl–Neelsen and periodic acid–Schiff (PAS) stains did not show any acid-fast bacilli or fungal organisms, respectively. PCR for mycobacterium gave a negative result.

What is your diagnosis?



Figure 1 Leonine facies, before treatment.

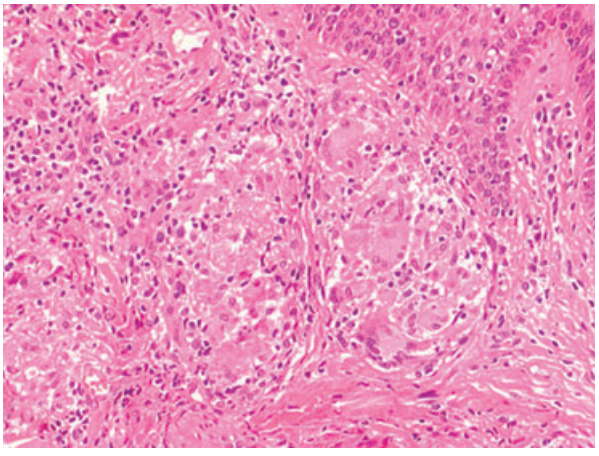


Figure 2 Well-circumscribed noncaseating epithelioid granulomas with Langerhans' giant cells (haematoxylin and eosin, original magnification × 200).

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Conflict of interest: none declared.

Accepted for publication 13 January 2008

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Diagnosis

Cutaneous sarcoidosis without systemic involvement.

Discussion

Cutaneous involvement in sarcoidosis may be classified as specific and nonspecific. Specific lesions are a result of granulomatous infiltrates, whereas granulomas are not found in nonspecific lesions. The morphology of the lesions includes papules, plaques, nodules, subcutaneous nodules, erythroderma, ulcerations, scars and alopecia. Follicular, verrucous, ichthyosiform, hypomelanotic, psoriasiform and annular lesions have also been described. Although the cutaneous lesions may present at any time during the course of the disease, they often present soon after disease onset. Cutaneous involvement occurs in 20–35% of patients with systemic sarcoidosis and may also occur in patients without systemic involvement.[1]

The histological differential diagnosis of sarcoidosis is comprehensive and includes the entire range of granulomatous reactions (tuberculoid leprosy, tuberculosis, leishmaniasis, syphilis, foreign-body reactions, granulomatous rosacea, granulomatous mycosis fungoides, cutaneous Crohn's disease, granuloma annulare, necrobiosis lipoidica and rheumatoid nodule),[1] but all of these diseases show significant histological differences from sarcoidal granulomas.

The histopathology of cutaneous sarcoidosis is characterized by naked tubercles, which are collections of epithelioid cells with a thin rim of lymphocytes. Caseation necrosis is not observed in sarcoid infiltration.[2] In granulomatous mycosis fungoides, epidermotropic infiltration is required for diagnosis.[3] Granulomas in cutaneous Crohn's disease usually have denser lymphocytic collections.[4] Tuberculoid leprosy has well-formed granulomas along neurovascular bundles with dense peripheral lymphocytic infiltrates. In lupus vulgaris, other histological features, in addition to caseation necrosis within the central portion of tubercles, denser collections of peripheral lymphocytes, and epidermal changes such as ulceration or pseudo epitheliomatous hyperplasia, usually permit its distinction from sarcoidosis.[2]

Ziehl–Neelsen and PAS stains should be performed to exclude the presence of acid-fast bacilli and fungal organisms. All granulomas should be examined with polarizing lenses to detect foreign bodies. Granulomas in necrobiosis lipoidica, granuloma annulare and rheumatoid nodule have a palisaded arrangement.[2] In granulomatous rosacea, dermal oedema with vascular dilatation and mixed inflammatory infiltration in vessels can be seen.[5]



Figure 3 Six months later the lesions had disappeared, after the patient had received low-dose systemic prednisone treatment.

Red–brown papules and plaques are the most common cutaneous findings of sarcoidosis.[1] Leonine facies is a rare presentation.[6]

Treatment options for sarcoidosis include strong topical or intralesional corticosteroids, methotrexate, chlorambucil, cyclophosphamide, azathioprine, thalidomide or psoralen ultraviolet A.[1]

We classified our patient as having cutaneous sarcoidosis without systemic involvement, based on the clinical, histopathological and laboratory findings. She was given oral prednisone 30 mg/day, gradually tapered from to 5 mg/day. The infiltration disappeared, and prednisone therapy was stopped after 6 months (Fig. 3).

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