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Mortimer's Malady revisited: A case of polymorphic cutaneous and systemic sarcoidosis

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ABSTRACT

Sarcoidosis is a systemic disorder with prominent cutaneous component. Skin lesions are of diverse morphology, of which few are specific for the disease. We describe a 30-year-old woman with polymorphic skin lesions including papules, plaques, and nodules, as well as uncommon variants like eyelid papules, palmar and digital nodules, tattoo sarcoid, as well as scar sarcoid. The patient also had stage II pulmonary sarcoidosis, and articular as well as reticulo-endothelial system involvement manifested by enlarged mediastinal and abdominal lymph nodes and hepatosplenomegaly. The presentation of polymorphic skin lesions with involvement of multiple extra-cutaneous systems is uncommon in a single patient.

Key words: Sarcoidosis, multisystem, sarcoid, tattoo, scar

INTRODUCTION

Sarcoidosis is a multisystem disorder of unknown etiology characterized by the presence of non-caseating granulomas in all affected organs. Jonathan Hutchinson in 1898 coined the term 'Mortimer's Malady' after the name of his patient Mrs. Mortimer who presented with multiple, raised, dusky-red, non-ulcerative and persistent patches, which Hutchinson considered different from tuberculous affliction.[1] Though other eponyms like 'Boeck's sarcoid' have also been in vogue, now the term 'sarcoidosis' is the most accepted for this condition. The clinical course of sarcoidosis is progressive with frequent remissions and relapses. Cutaneous lesions have been classified as specific or non-specific.[2] Non-specific skin lesions like erythema nodosum are often associated with an acute presentation and generally portend a good prognosis. Specific skin lesions including papules, plaques, nodules, lupus pernio and scar infiltration among others, tend to be chronic and require therapy for resolution.[3,4] Sarcoidosis frequently involves the lungs, lymph nodes, liver, spleen and eyes. We present a case of a young female patient with presence of polymorphic lesions including tattoo and scar sarcoidosis as well as widespread involvement of lungs, joints, liver, spleen and mediastinal as well as abdominal lymph nodes.

CASE REPORT

A 30-year-old female patient presented with multiple red raised asymptomatic lesions starting from the trunk and followed by involvement of the face, limbs and palms and soles within a period of 6 months. She complained of persistent dry cough and exertional dyspnoea for more than a year, associated with malaise, pain and morning stiffness in joints of hands and feet. Her past medical history, personal, occupational and family histories were insignificant. Cutaneous examination revealed presence of multiple erythematous infiltrated papules and plaques over the trunk, thighs and legs and erythematous infiltrated nodules over forehead and nose. Multiple erythematous nodules were also noticed over palms and the tips of fingers and distal plantar aspect of toes [Figure 1]. Upper eyelids had small, erythematous infiltrated papules [Figure 2]. A close look at 15-year old black-pigment tattoo over left forearm disclosed infiltration with tiny erythematous papules [Figure 3].

Skin biopsy from a papule on the knee revealed confluent non-caseating granulomas with epithelioid cells and few lymphocytes, suggestive of cutaneous

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sarcoidosis. Biopsy from the tattoo papule also showed similar dermal infiltrate with presence of macrophages laden with engulfed tattoo pigment, both inside and surrounding the granulomas [Figure 4]. Interestingly, following the skin biopsy from the knee, a nodular lesion rapidly developed over the scar [Figure 5]. Peripheral lymph nodes were not enlarged. Clinical examination of cardiovascular, respiratory and neurological systems was normal. Abdominal assessment revealed significant hepatosplenomegaly. Tenderness and mild stiffness were noticed over metacarpo-phalangeal and interphalangeal joints of hands and feet, without any swelling or deformity.

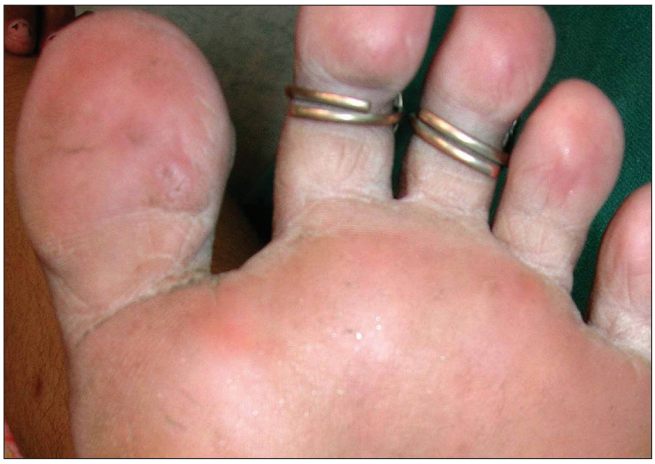

Figure 1: Erythematous nodules over distal plantar aspect of toes

The hematological and bio-chemical tests including liver and kidney function tests and serum calcium and phosphate were normal except for elevated erythrocyte sedimentation rate (40 mm in first hour). Serum levels of angiotensin converting enzyme were elevated (140 U/L). Sputum examination and culture for mycobacteria excluded tuberculosis. Tuberculin skin test (5 Tuberculin units) was negative. ELISA for

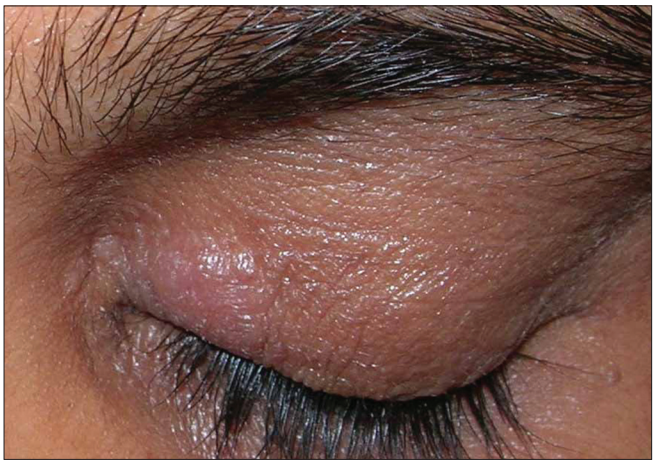

Figure 2: Eyelid papules

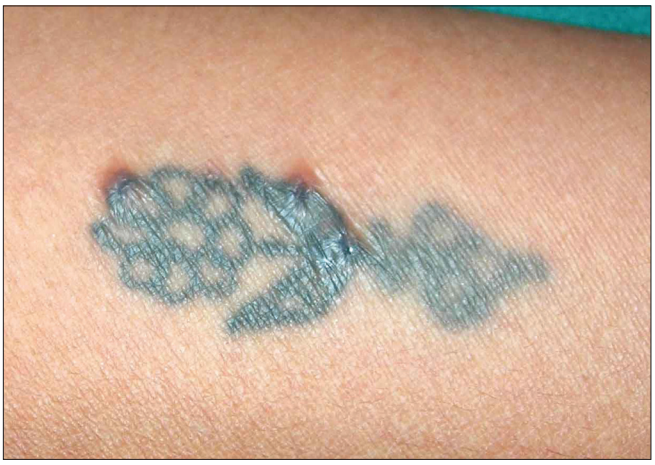

Figure 3: Erythematous papules infiltrating an old tattoo over forearm

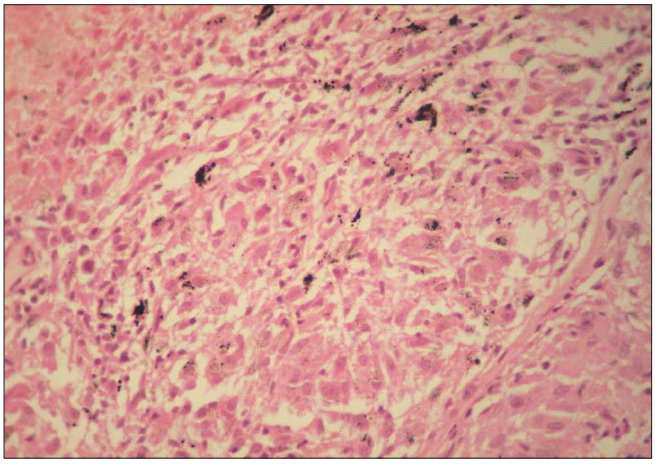

Figure 4: Non-caseating granulomas in dermis with engulfed tattoo pigment inside and surrounding the granulomas (H&E, ×200)

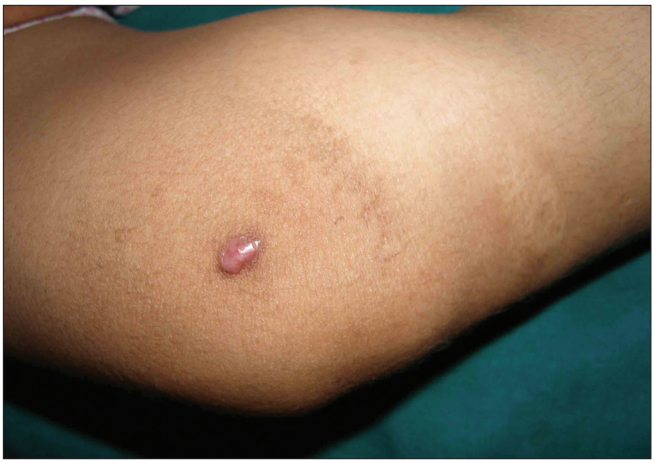

Figure 5: Scar sarcoidosis

HIV was also negative. Chest X-ray showed bilateral hilar enlargement with patchy consolidation in right middle lobe. Contrast enhanced computed tomography (CECT) of chest revealed markedly enlarged lymph nodes in the mediastinum, involving pretracheal, right paratracheal, subcarinal and carinal regions and in bilateral lung hila. Patchy alveolar consolidation with evidence of early fibrosis was seen in right middle lobe. These changes were consistent with early stage II of pulmonary sarcoidosis. Spirometry suggested moderate restrictive lung disease. Abdominal ultrasound scan revealed hepatomegaly (17 cm) and splenomegaly (15 cm). These were confirmed by abdominal CECT scan which additionally showed the presence of multiple enlarged lymph nodes in the portal, peripancreatic, and para-aortic regions and splenic hilum. Juxta-articular osteopenia was noted on X-rays of hands and feet, though no lytic lesion was visualized. The electrocardiogram and echocardiogram were normal. Patient was started on oral prednisolone 40 mg and hydroxychloroquin 200 mg twice a day. Treatment was followed by prompt and marked improvement in malaise, arthralgia and pulmonary symptoms. Skin lesions also subsided within 4 weeks with atrophic scarring developing at few sites.

DISCUSSION

Cutaneous involvement is known to occur in 20-35% cases of systemic sarcoidosis.[2] Our patient had specific sarcoidal lesions like papules and plaques with eyelid papules and nodules over palms and digital tips, which are uncommon lesions, reported in isolated cases previously.[5,6] The involvement of an old tattoo with infiltrated papules is another rare and notable finding. Following the first report by Madden,[7] tattoo sarcoid has been reported infrequently.[8] Tattoo sarcoid occurs in two different clinical settings. A tattoo may become involved after several years as part of idiopathic sarcoidosis, usually in absence of other cutaneous or systemic lesions. Alternatively, the tattoo pigment may elicit granulomatous hypersensitivity and induce systemic sarcoidosis.[8,9] In the latter situation, pigment particles are frequently detected within granulomas of tattoo sarcoid as well as non-tattoo skin lesions. The case presented here is a patient with true idiopathic sarcoidosis rather than pigment-induced disease owing to the long time span between the engraving of tattoo and appearance of skin lesions and the absence of pigment in granulomas from the knee. Scar sarcoidosis is another rare but specific form of the disease, reported in 2.9% to 13.8% cases.[4,10] It may occur at a scar resulting from biopsy, burns, surgery, herpes zoster or BCG vaccination. In a series of 23 cases of cutaneous sarcoidosis from India, Mahajan *et al* described two patients with sarcoid nodules developing over scars of childhood injuries.[11] Infiltration of a recent biopsy scar in a suspected or known case of sarcoidosis indicates disease activity, as evident in this patient.[10]

Our patient had stage II pulmonary disease, arthralgia and reticulo-endothelial system involvement in the form of hepatosplenomegaly and enlarged mediastinal and abdominal lymph nodes. Since these were established by non-invasive radiological investigations, we did not subject the patient to procedures like mediastinal node biopsy or transbronchial lung biopsy for diagnosis. A few other investigations helpful in the diagnosis of sarcoidosis deserve mention. Bronchoalveolar lavage (BAL) is a minimally-invasive and safe technique for the diagnosis of sarcoidosis. A high CD4/CD8 lymphocyte ratio (greater than 4.0) in the BAL fluid differentiates sarcoidosis from other interstitial lung diseases. The sensitivity of this test is variable (42% to 59%) and 10% cases of sarcoidosis are known to have low CD4/CD8 ratios.[12] Whole body gallium-67 scan aids in delineation of the extent of the disease, detection of hitherto unsuspected lesions (for example, salivary glands, spleen, etc.), differentiation of granulomatous thoracic lesions from fibrosis and is considered to be a sensitive indicator of disease activity.[13] The shortcomings of gallium scan include the risk of radiation exposure and the high cost of investigation. Thus, in select cases, investigations like BAL and gallium-67 scan may be useful in diagnosis and monitoring of disease activity in sarcoidosis.

The presence of such diverse types of cutaneous lesions as well as multisystem involvement of sarcoidosis in a single patient is unusual and infrequently reported. Sharvill D described the concomitant presence of cutaneous, conjunctival and mediastinal sarcoidosis.[14] Cerri *et al*. documented a 62-year-old woman with cutaneous, pulmonary, ocular, cardiac, bone and articular involvement.[15] A recent case described the presence of erythema nodosum, annular plaques, and subcutaneous nodules with pulmonary, cardiac, ocular and bone involvement in a 56-year-old woman.[16]

Thus, this case highlights that sarcoid is a great imitator

in dermatology and skin lesions often represent just the tip of the iceberg.

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