

Sarcoidosis with Skin Manifestations - Two Case Reports and Review of Literature

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Sarcoidosis is a granulomatous disease of unknown etiology involving various organ systems. The skin involvement includes a variety of lesions. In this report two cases of sarcoidosis with different skin manifestations are described.

Introduction

Sarcoidosis is a multi-system granulomatous disease of unknown etiology, most commonly affecting young adults and presenting most frequently with bilateral hilar lymphadenopathy, pulmonary infiltration, eye or skin lesions¹. Cutaneous lesions are present in 20-35% of patients and are polymorphous². The diagnosis is established most securely when clinico-radiographic findings are supported by histologic evidence of non-caseating granulomas. Here we report two cases of sarcoidosis with skin involvement.

Case 1

A 34 years old male presented with a two years history of multiple skin lesions all over the body. The lesions consisted of multiple erythematous papules and plaques on back and chest with scaling and some indurated areas (Figures 1 and 2).



Figure 1. A patient with erythematous plaques on the back.



Figure 2. Magnified view of the lesions shown in Figure 1.

A skin biopsy revealed non-cascating granulomas and the patient was started on anti-tuberculous therapy, which he continued for seven months, but new lesions kept on appearing. He came to this hospital for second opinion. Besides skin lesions he gave a history of recent weight loss and low grade fever. General examination apart from skin lesions was unremarkable with no lymphadenopathy. Respiratory system examination was also normal. There were no other systemic findings, in particular liver and spleen were not palpable. His investigations revealed an ESR of 9 mm/hour, serum calcium of 8.9 mg%, angiotensin converting enzyme levels of 135 IU/L (normal 8-52 IU/L). His mantoux test was negative. Skin biopsy was repeated which once again showed non-caseating granulomas. Chest X-ray showed lymphadenopathy involving the mediastinum and paratracheal region. CT revealed bilateral mediastinal and paratracheallymphadenopathy. A repeat biopsy again showed non- caseating granulomatous inflammation without necrosis (Figure 3).

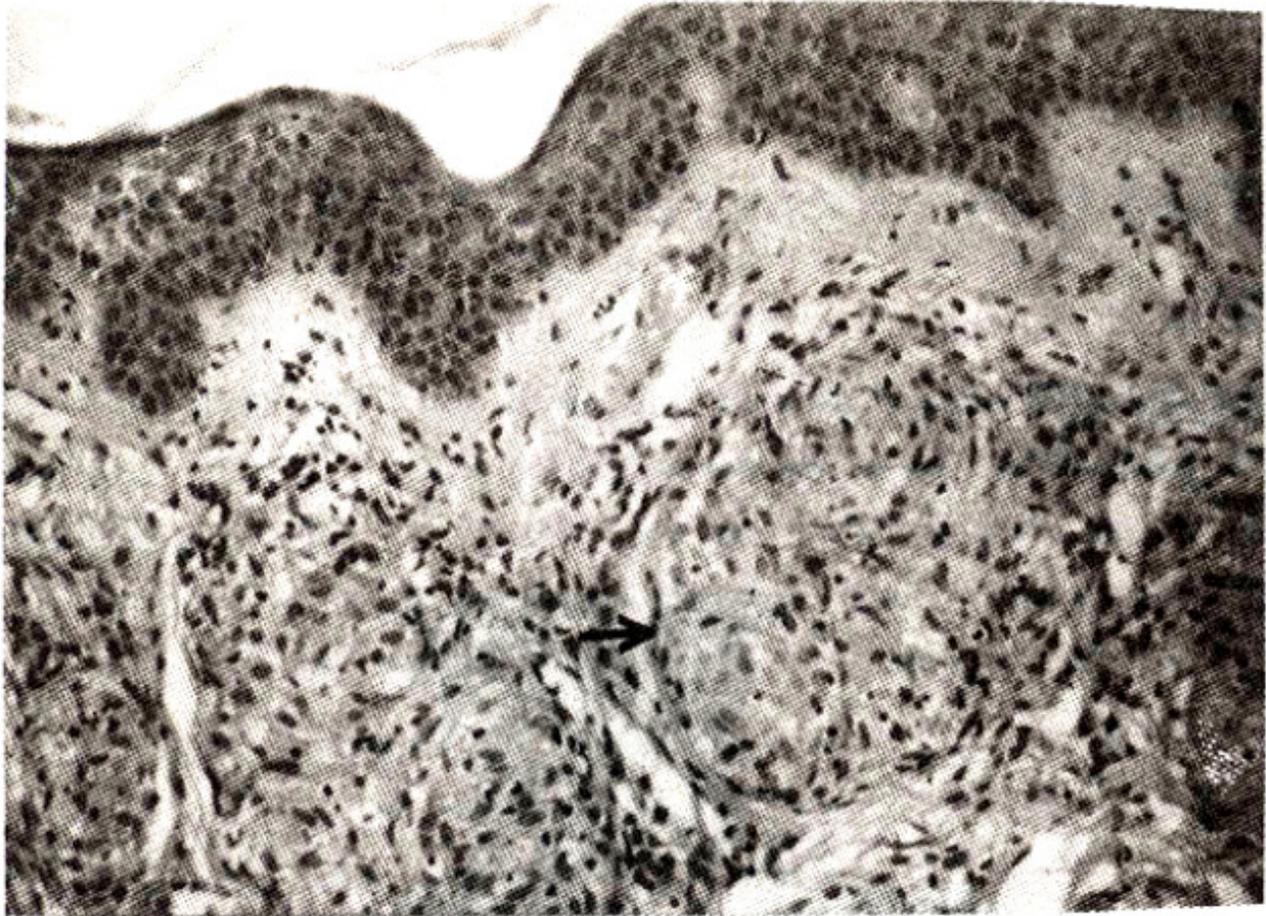


Figure 3. Large epitheloid cell granuloma (arrow).

The patient was started on topical steroids, but because of the extensiveness of the lesions and lack of substantial improvement he was started on oral prednisolone 20mg bid. On follow-up remarkable improvement of the lesions was noted and the patient was continued on same treatment with a plan for gradual tapering of the steroids.

Case 2

A 45 years old male originally presented in 1992 with complaints of dry cough and dyspnea. Chest x-ray revealed bilateral hilar lymphadenopathy and the CT scan showed bilateral hilar and mediastinal lymph node enlargement. He was advised to have a lymph node biopsy but he did not agree and was lost to follow-up. The patient again presented two years later with complaints of dry cough, progressively increasing dyspnea and weight loss. In the course of time he had also developed some skin lesions. His physical examination revealed presence of subcutaneous nodules on the back. Nodules were 0.5 to 1 cm in size, pale in colour and non-tender. They were scattered all over the back (Figure 4),

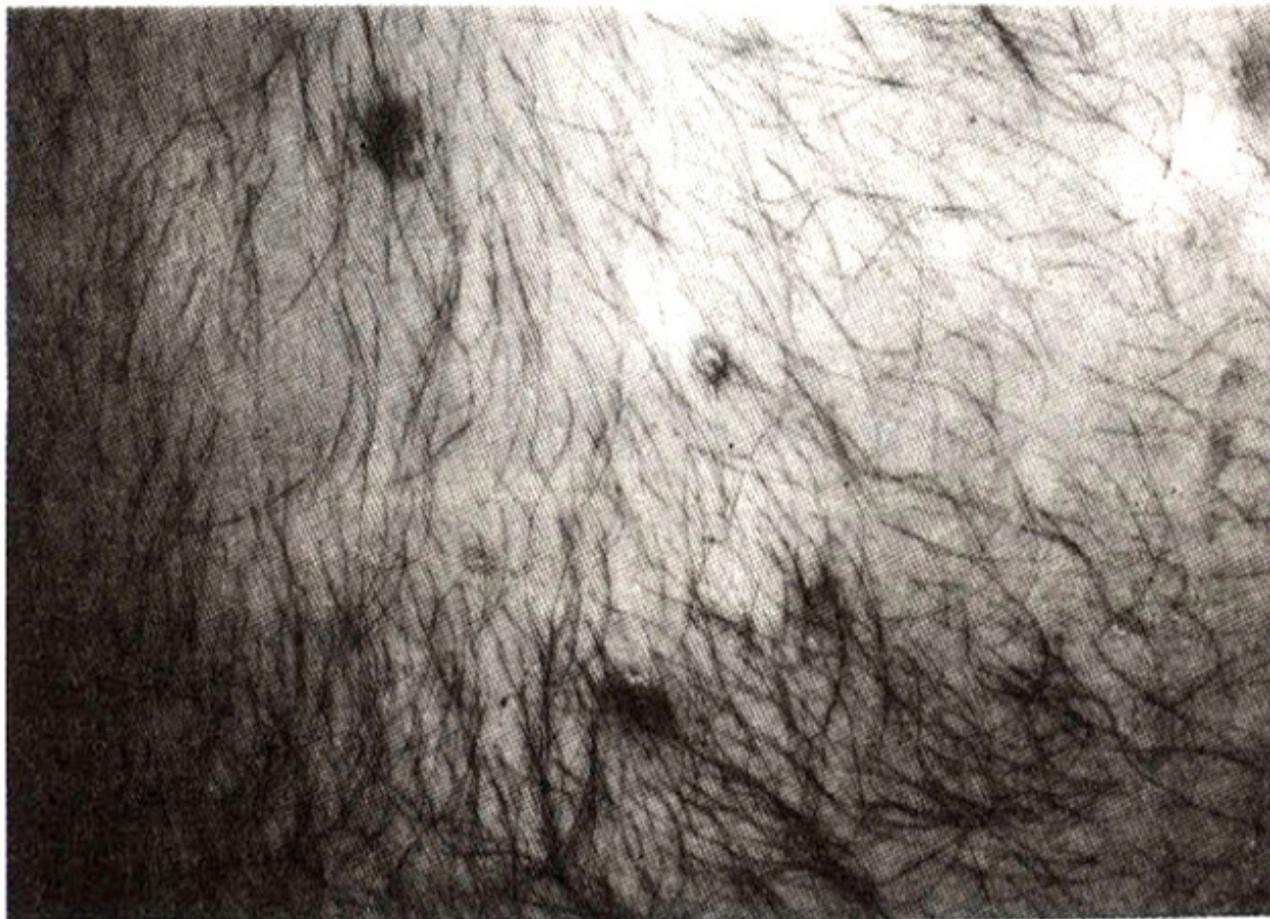


Figure 4. Subcutaneous nodules of sarcoidosis.

there was no hepatosplenomegaly or any other findings and rest of systemic examination was normal. The investigations showed an ESR of 51 mm/hour, ACE levels of 91.5 IU/L (normal 8-52 IU/L), serum Ca²⁺ of 10.3 mg% and mantoux test was negative. A chest X-ray done in 1994 showed hilar lymphadenopathy and reticular pattern at the bases. Biopsy of the skin lesions showed non-caseating granulomas. He was started on oral steroids 40 mg OD. On reviewing the patient few weeks later considerable improvement was seen in his skin lesions. His dyspnoea and cough also showed marked improvement.

Discussion

The skin involvement in sarcoidosis is polymorphous and the lesions may be specific or non-specific. The important specific lesions are lupus pernio, plaques and maculopapular eruptions. The important non-specific lesion is erythema nodosum. Other skin lesions include alopecia, erythrodermas, subcutaneous nodules, erythema multiforme, ichthyosis, dystrophic calcifications and verrucous outgrowths. Nail involvement is rare in sarcoidosis³. Lupus pernio, the most characteristic of all sarcoid lesions, is a chronic, violaceous, indurated skin lesion with a predilection for the nose, cheeks, ears and lips. It is more common in the females and those over forty. Compared with sarcoidosis overall, lupus pernio has a closer affinity with sarcoidosis of the upper respiratory tract, bone cysts and lacrimal gland involvement¹.

The maculopapular lesions are the most common skin manifestations of sarcoidosis in the black

patients. The waxy translucent lesions with a distinct flat top vary from 2-6 mm in diameter. They characteristically occur on the face, lids, around the orbits, in the nasolabial folds and on the nape and upper back³. Plaques of sarcoidosis usually manifest as elevated, indurated purplish patches commonly located on the limbs, face, back and buttocks. The distribution is usually bilateral and symmetrical. The central part of the plaque is pale and atrophic, the periphery indurated elevated and dark. In the presence of telangiectatic vessels the lesions are called angioliupoid. Subcutaneous nodules, also called Darier-Roussy sarcoidosis are oval, firm, painless structures that arise deep in the dermis and subcutaneous tissue of the trunk and extremities. Scars from atrophy, trauma, surgery or venipuncture may become purple, swollen and tender either at the time the patient presents or during reactivation of the disease⁴. Ulcerative sarcoidosis is rare, occurs more frequently in black women and usually involves the legs. Erythema nodosum is a hypersensitivity reaction that is the most common non-specific cutaneous manifestation of sarcoidosis. Systemic manifestations such as fever, malaise and polyarthralgia occur in about fifty percent of patients with erythema nodosum⁵. Because of the high rate of spontaneous resolution, they seldom require steroid therapy. The exact prevalence of sarcoidosis in Pakistan is not known. It is probably underdiagnosed and being overshadowed by other granulomatous diseases like tuberculosis. The criteria for establishing the diagnosis of sarcoidosis includes (1) a compatible clinical or radiological picture or both; (2) histological evidence of non-caseating granuloma and (3) negative special stains and cultures for other entities (e.g., acid fast bacilli or fungi in sputum or tissue specimen)³. The serum calcium, ESR and ACE levels are elevated significantly in patients with sarcoidosis and serial measurements may prove useful in the monitoring of these patients, but their value in the diagnosis of sarcoidosis is limited as they are non-specific. The definitive diagnosis still requires the demonstration of non-caseating granuloma in the involved tissue¹. The management of cutaneous sarcoidosis must take into account the seriousness of the accompanying internal involvement and the natural history of the particular type of lesion. Papular lesions are likely to fade without treatment, whereas lupus pernio does not. High potency topical steroids may sometimes prove helpful, as may intralesional triamcinolone injections. Cryotherapy and radiotherapy have occasionally been used. In systemic therapy at present, corticosteroids are the most effective treatment, given at first at a relatively higher dose and then tapering over a period of several weeks to a lower maintenance dose on alternate days⁶. Cytostatic drugs may be tried if corticosteroids are contraindicated or have been ineffective. Methotrexate seems, so far, to have the best track record in the treatment of cutaneous as well as disseminated sarcoidosis. Chlorambucil and azathioprine may be used for their steroid sparing effect⁷. Other drugs which have been tried with some success include allopurinol⁸, antimalarials¹⁰ and oxyphenbutazone¹⁰. The chance of spontaneous remission favours a conservative approach to systemic therapy, which will usually carry the hazards of long term immunosuppression. At any time the pattern of the disease may change, but an expectant policy is best if the course is not progressive and if vital structures are not involved. The two cases discussed reiterate that sarcoidosis with its various clinical manifestations exists in Pakistan and poses a difficulty for the physician in diagnosis and management.

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