Case Report

Cutaneous Manifestations of Sarcoidosis : Reports of Two Cases

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Abstract:

Sarcoidosis is a multisystem disease that may involve any organ or system. Cutaneous manifestations occur in 25- 30% of patients with sarcoidosis. Sarcoidosis can present with reactive non- specific lesions like erythema nodosum or specific manifestations showing granulomatous microscopic pathology including papules, plaques, lupus pernio, scar

sarcoidosis, and rare morphologies such as alopecia, hypopigmented patches and ichthyosiform lesions. Here we present two interesting cases who presented with cutaneous manifestations and causing diagnostic diemma.

Key words: *Sarcoidosis, Cutaneous manifestations, Bangladesh.*

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Introduction

Sarcoidosis is a chronic inflammatory multisystem granulomatous condition of obscure etiology. The lungs are the most frequently affected organ , but the skin, eyes, liver, lymph nodes, bones, kidneys, parotid glands are also commonly involved. Skin involvement occurs in approximately 25% of cases[1]. In about 10% case skin is the solely affected organ[2]. The lesions may be verrucous, ichthyosiform, hypomelanotic, psoriasiform or alopecic. Papules of sarcoidosis may be of various colors, including red, reddish-brown, violaceous, translucent, or hyperpigmented[3]. Sarcoid lesions are asymptomatic but approximately 10-15% may itch[2]. Erythema nodosum is the most common nonspecific cutaneous finding in sarcoidosis. Because lesions assume a vast array of morphologies, cutaneous sarcoidosis is known as one of the "great imitators" in dermatology. Here we report two cases of sarcoidosis with cutaneous manifestations which put us to diagnostic challenge.

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Case 1

A 30- year-old lady presented with erythematous patches on both legs of 8 months duration. These were sometimes associated with slight burning sensation. Patches were increasing in

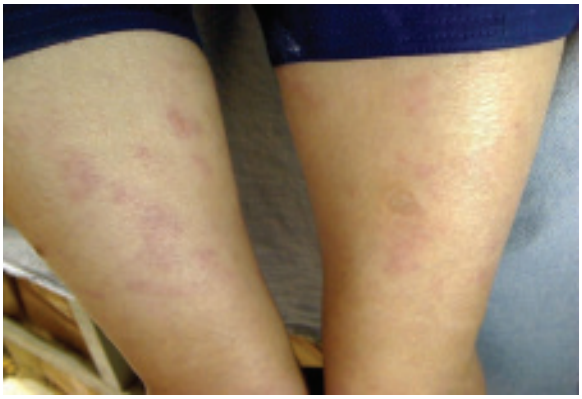

Figure 1: *Erythematous patches on anterior surfaces of both legs*

size and number. There was no associated history of sore throat, joint pain, chest pain, fever or weakness. For this, she visited several physicians and was treated with different topical steroid and antibacterial agents without any improvement.

On examination, there were multiple erythematous macules and patches of various sizes and shapes distributed on extensor surfaces of both legs. Lesions

were nontender, local temperature and sensation were normal. Diascopy revealed no colour change of the lesions. Nail, hair and mucosa were normal. Other systemic examinations revealed no abnormality.

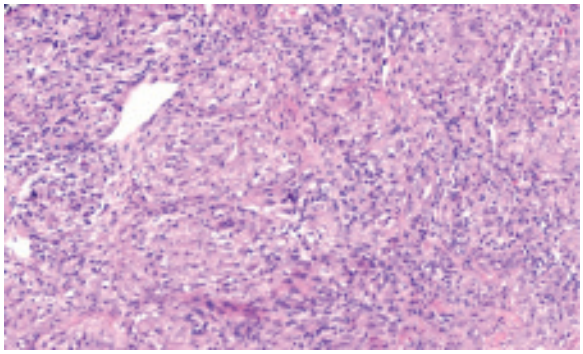

Figure-2: *Skin biopsy showing noncaseating naked granulomas*

Routine hematologic and urine investigations were normal. Serum Angiotensin converting enzyme (ACE) titer was 80 U/L. Serum calcium, Serum total protein, liver function tests (LFT), renal function tests (RFT), ultrasonogram (USG) abdomen were normal. Mantoux test (MT) was 0 mm.

Skin biopsy for histopathology revealed large number of non caseating granulomas replete with aggregates of epithelioid cells and few giant cells. The granulomas were mostly present in reticular dermis involving adnexal structures and fat. The granulomas were naked with lyphocytes around the granulomas. The epidermis was free and no acid fast bacillus (AFB) was seen. X-ray chest postero-anterior (P/A) view showed bilateral hilar and paratracheal lymphadenopathy. Pulmonary function test revealed mild restrictive abnormality. Computed tomography (CT) (multi slice) of chest showed enlarged mediastinal and bilateral hilar lymph nodes. CT guided fine needle aspiration cytology (FNAC) showed moderate cellular material containing lymphocytes, polymorphs, pulmonary macrophages, a few giant cells and focal collections of epithelioid cells. No caseation necrosis is seen. No malignant cell is seen either. Eletrocardiogram (ECG) was within normal limit. Echocardiogram revealed no wall motion abnormality. Normal LV systolic function (EF: 61%).

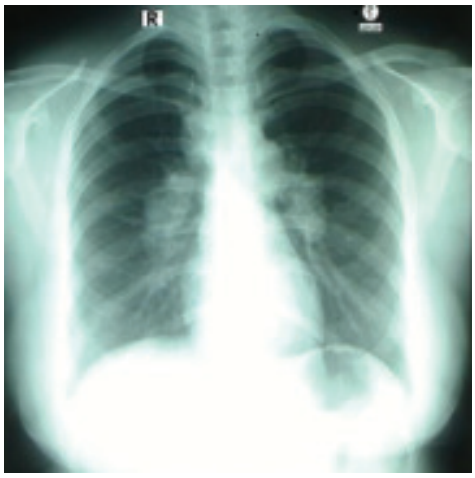

Figure-3: *X-ray chest showing bilateral hilar and paratracheal lymphadenopathy*

We treated her with tablet Prednisolone 30 mg /day as starting dose and tapering over 8 weeks period and tablet hydroxychloroquine sulphate 200 mg once daily . Complete resolution of cutaneous and radiological finding occurred within three months.

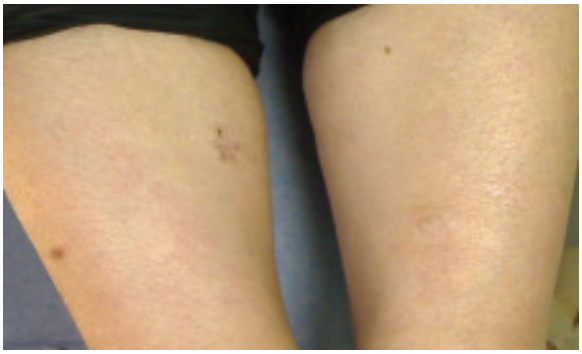

Figure-4: *Resolution of skin lesions after 3 months treatment*

Case 2

A 32- year-old lady presented with an erythematous plaque on forehead for one and half year. Initially it was small but gradually increasing in size. The lesion was slightly painful. Therewas no associated joint pain, fever, chest pain or weakness. For this, she visited different dermatologists and was treated as cutaneous tuberculosis and was treated with anti TB drugs for four months but condition did not improve. On examination, there was an erythematous infiltrated plaque with irregular suface and margin on forehead. It was slightly tender with normal local temparature and sensation. Diascopic examination revealed apple jelly color change of the lesion. Other examinations revealed nothing abnormality.

Skin biopsy showed a granulomatous reaction pattern characterised by multiple discrete predominantly epithelioid granulomas. Necrosis is absent. The granulomas are typically 'naked' with few surrounding lymphocytes and a rim of mild dermal fibrosis.

X ray chest P/A view showed bilateral hilar lymphadenopathy. Pulmonary function test was Within normal limit, MT was o mm. ECG was Normal. Other hematological and biochemical parameters were within normal limit.

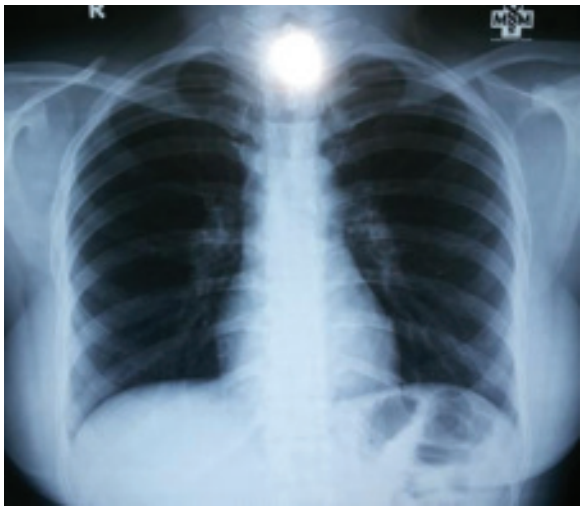

Figure-5:

We treated her with tablet prednisolone 30 mg /day as initial dose and tapering over a period of 12 weeks and tablet Hydroxychloroquine sulphate 200 mg once daily for 12 weeks. Near- complete resolution occurred after three months.

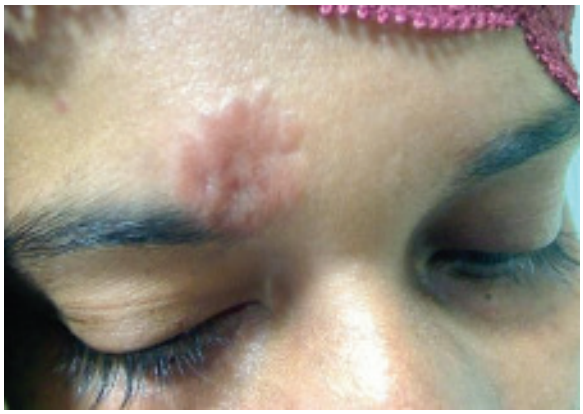

Figure-6: *Erythematous indurated plaque on forehead*

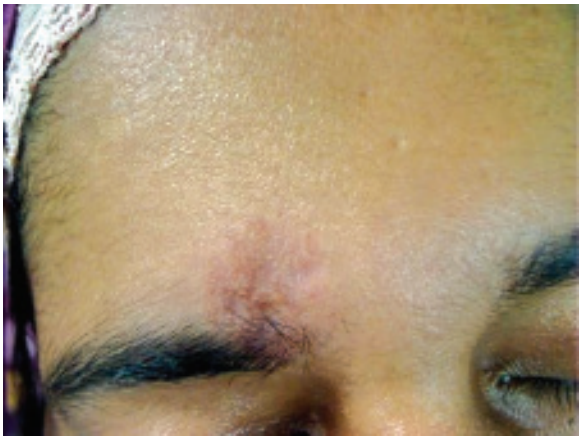

Figure-7:

Discussion

The first descriptions of sarcoidosis in the late 1800s were limited to its skin manifestations[4]. The term sarcoidosis is derived from pioneer norweigian dermatologist Caesar Boeck's 1899 report of what he described as "multiple benign sarkoid of the skin," because he believed the lesions resembled sarcomas but were benign[5]. The disease usually begins at around 40 years of age and nearly two-thirds of the cases are females[6]. It occurs worldwide with the highest prevalence in Scandinavia and the prevalence was 20/ 100,000 in 1975 but recent report says it to be 64/ 100,000[2]. In Bangladesh, there is not much statistical data regarding the prevalence or pattern of sarcoidosis. Because of rarity of this disease, higher incidence of skin TB in this country and inexperience of some cytopathologists, many cases clinically consistent with sarcoidosis are falsely labeled as tuberculosis. We believe that sarcoidosis is more prevalent in Bangladesh than currently is predicted.

A variety of candidate antigens have been suggested as provocative agents for the cascade of immunologic events that eventuate in sarcoidosis. Infectious agents such as Mycobacteria, Propioni-bacterium acnes and Chlamydia have been associated with sarcoidosis. Mineral dusts, such as silica, iron, and titanium, Combustible wood products. Genetics play a major role in determining susceptibility to sarcoidosis.

In Bangladesh two case reports on Lofgren syndrome, one case report of lupus pernio and one case of maculo-papular eruption were previously reported[7,9]. Among the numerous reportedmorphologic presentations are

papules, micropapules, plaques, subcutaneous nodules, scar sarcoidosis, lupus pernio, erythema nodosum, ulcer and alopecia[10]. Rare presentations like follicular, verrucous, ichthyosiform, hypomelanotic, and annular lesions have also been described[11]. In India lichenoid and psoriasiform pattern of sarcoidosis have been described in a case report[12]. Morpheaform sarcoidoisis has been reported in one case report[13].

Asymptomatic bilateral hilar adenopathy on chest radiograph almost always represents sarcoidosis. It has been suggested that histologic confirmation of sarcoidosis may not be required in asymptomatic patients with bilateral hilar adenopathy, provided the physical examination, complete blood cell count, and routine blood tests are all normal and there is no prior history of malignancy. Although a confirmed diagnosis of sarcoidosis requires proof of granulomatous involvement in at least two separate organs, histologic confirmation is usually not required in the second organ[5]. In sarcoidosis MT is negative and in TB it is usually positive. Inappropriate treatment with anti-TB chemotherapy may give rise to multi drug resistant-TB (MDR-TB). In both of our cases we found cutaneous noncaseating granuloma on skin biopsy, asymptomatic bilateral hilar lymphadenopathy and negative MT. Early diagnosis is possible by doing skin biopsy and it may help to avoid misdiagnosis and can ensure appropriate treatment of these cases.

Conclusion

Recognition of cutaneous lesions is important because they provide a visible clue to the diagnosis and are an easily accessible source of tissue for histologic examination. Any indurated chronic asymptomatic lesion of skin should raise suspicion of other common granulomatous diseases like sarcoidosis.

Conflict of interest: None

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