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Case Report of a Rare Cutaneous Manifestation of Sarcoidosis

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Sarcoidosis is a multisystem granulomatous disease of unknown etiology and pathogenesis with a chronic course characterized by multiple remissions and relapses. Besides a brief review of relevant literature, this paper reports on a 70-year-old Saudi male patient who presented with a rare form of cutaneous sarcoidosis masquerading as psoriasis. Physical examination revealed presence of multiple, symmetrical erythematous plaques covered with silvery white, adherent scales involving elbows, knees, scalp and side of both buttocks but complete absence of similar lesions in genitalia and nails. Though radiological findings suggested sarcoidosis, cutaneous sarcoidosis was finally diagnosed by morphological changes-noncaseating, granulomatous inflammation as revealed by skin biopsy. After two months of treatment with prednisolone and other medications, this patient showed considerable improvement and all lesions disappeared. In the light of this case and review of literature, several aspects of cutaneous sarcoidosis presenting as psoriasis are discussed.

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Sarcoidosis, a systemic granulomatous disease of undetermined etiology and pathogenesis, involves different body organs with a persistent course often characterized by remissions and relapses. This is a multi-organ disease that may involve lungs, lymph nodes, eyes, myocardium, kidneys, spleen, liver and central nervous system. Notably, skin alone is affected in about 9% to 37% of cases[1]. Unlike western world, there is scanty epidemiological data on systemic and cutaneous sarcoidosis in Arabian Gulf region[2-4]. By and large, many cutaneous descriptions, form common to very rare, of sarcoidosis are reported in the literature[4]. Although several laboratory abnormalities may be found, no single diagnostic test for sarcoidosis is available[5]. The cutaneous manifestations of sarcoidosis often help dermatologists to diagnose its systemic form[6].

This case report highlights a very rare presentation of cutaneous sarcoidosis with systemic involvement.

THE CASE

A 70-year-old, non-smoker Saudi male patient with a history of 5 months cough and shortness of breath but no complains of orthopnea or paroxysmal nocturnal dyspnea was

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admitted to King Khalid University Hospital, Riyadh, Saudi Arabia. A systemic review of history revealed no other symptoms. As an incidental finding during general physical examination, skin lesions of 15 years duration were observed on several body areas. Family history was negative for similar skin lesions. The patient was referred to dermatological services for further evaluation. Skin examination revealed multiple, symmetrical erythematous plaques covered with silvery white, adherent scales involving elbows, knees, scalp and side of both buttocks (Fig 1 a & b). The genitalia and nails were found to be normal. A batter of laboratory investigations including CBC, FBS, LFT and lipid profile showed no abnormal values and moreover PO2 and PCO2 were 70 and 49 with 94% saturation. Chest X-Ray showed bilateral diffuse fine reticulo-nodular pattern with chronic interstitial lung disease (Fig 2. a) whereas CT scan showed interstitial pulmonary fibrosis (Fig.2 b). Bronchoscopy was normal except for superficial generalized inflammation. Skin biopsy showed noncaseating, granulomatous inflammation (Fig. 3). Acid fast bacilli (AFB) do not exist in the fite stain. No foreign material was seen under polarized light examination. All these findings were consistent with sarcoidosis. The patient was treated with prednisolone 20mg qid, elocume cream and salisalic acid 10 % in vaseline topically. Further, he was advised to use sebolux shampoo for scalp lesions. Within 2 months, the patients showed substantial improvement with the disappearance of all lesions. Prednisolone was then tapered to 15mg and emollint was added. Since then, the patient did not follow hospital clinics.

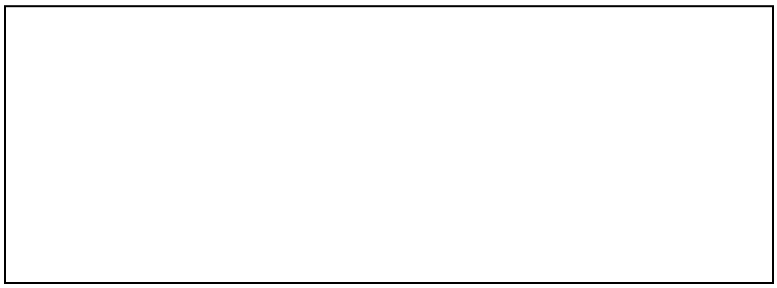

Figure 1. a- Well-defined plaque covered with adherent scales over elbows.

Figure 1. b- Similar lesion at side of his buttock.

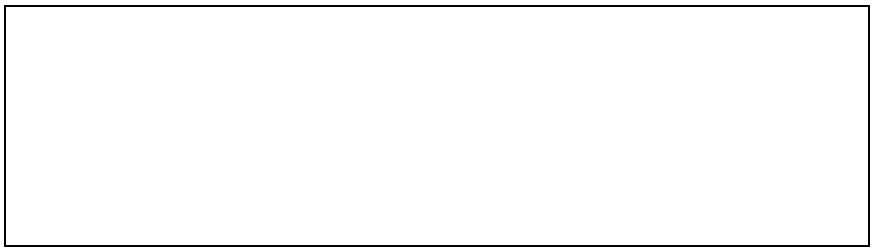

Figure 2. a- Chest X-ray AP revealed diffuse coarse reticulo-nodular pattern

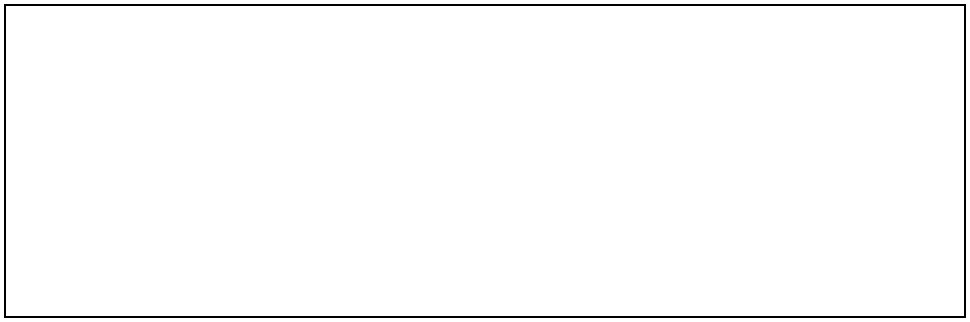

Figure 2. b- CT-Scan showing same finding mainly at periphery indicating interstitial fibrosis.

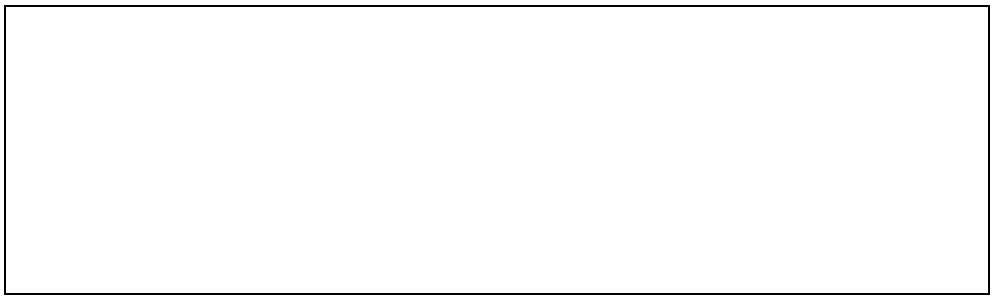

Figure 3. Photomicrograph showing well formed non-caseating granulomatous reaction

DISCUSSION

Reportedly, cutaneous manifestations in sarcoidosis occur in up to one third of patients who are known to suffer from systemic sarcoidosis. This is often classified as "specific" type with granulomatous reaction demonstrated in skin biopsy and alternatively "non-specific" type is characterized mainly by reactive changes but no traces of granulomas[7]. In about 33% of cases, skin lesions precede the systemic disease while in the remaining the skin manifestations appear simultaneously[8].

Reportedly, the incidence of sarcoidosis varies globally[4]. There are no prospective studies that have addressed the exact prevalence of sarcoidosis in the Arabian Gulf countries. However, some retrospective reports have projected some figures about sarcoidosis[9-11].

Samman et al carried out a retrospective review of charts of all patients with confirmed diagnosis of sarcoidosis in a general hospital in Western Saudi Arabia over a period of 11 years[11]. Seventeen of their reviewed cases were native Saudis and on comparison of their results with western world pattern of this disease, thy found that females were preferentially afflicted by this disease. Moreover, they discovered that cardiac system, eye, parotid and CNS were uncommonly involved among their population study[11]. However, another study comprising of 20 sarcoidosis cases revealed relatively frequent eye involvement as compared to western pattern of the disease[10]. Moreover, they found more severe constitutional symptoms in their reviewed cases than it has been reported in the literature, which is concordant with a study from Kuwait[9]. Unsurprisingly, none of these studies has reported a psoriasis-like presentation of sarcoidosis. Notably, as these studies consisted of small samples with retrospective design, their results and figures may not be generalized.

Many morphologic skin lesion types have been described in sarcoidosis. Like syphilis, it is considered as one of the "great imitators" in dermatology. Common granulomatous skin lesions in sarcoidosis, as also revealed in this case, include macules, papules, nodules, plaques, subcutaneous nodules, lupus pernio, and infiltrative scars[4,8]. Among all lesions, papules are considered as the most common of the specific cutaneous lesions while erythema nodosum is the most common nonspecific cutaneous lesion of sarcoidosis and has been reported in 25% of cases[4,12,13]. Moreover, lupus pernio is the most characteristic cutaneous lesion of sarcoidosis and is the most common in African-American women[4,6,8]. There are other less common, specific presentations including specific and nonspecific nail changes of sarcoidosis that have been reported in the dermatological literature[4]. Specific papulo-sqaumous skin eruptions are also rare presentation in sarcoidosis. Many reported cases of sarcoidosis, mostly from nonwhite patients (75%), showed ichthyosiform morphology and eruptions commonly on legs[14,15]. It is wise to note that despite reports of rare manifestations of sarcoidosis, there is no detailed description of psoriasis-like lesions for cutaneous sarcoidosis in the literature. Cutaneous lesions of sarcoidosis except erythema nodosum and lupus pernio are known to have no prognostic significance or correlation with disease severity or systemic involvement[6,8,16]. Erythema nodosum, although non-specific, is the hallmark of acute sarcoidosis with spontaneously resolving course and tends to have a good prognosis[17,18]. Lupus pernio, a sarcoid specific, has been associated with bone cysts, sarcoidosis of the upper respiratory tract, and pulmonary fibrosis and heralds bad prognosis[16,19].

To accurately diagnose sarcoidosis is a challenge for physicians as no single laboratory test can clinch the correct diagnosis[5]. Like this patient, cases are accurately diagnosed with sarcoidosis only when a set of clinical, supportive radiological pictures, and histological findings of noncaseating granulomas are available, the latter evidence excludes other potential causes, such as infections[5,20]. Recognition of cutaneous lesions of sarcoidosis is important because they provide a visible clue to support the diagnosis and an easily accessible source of tissue for histological examination. Punch or incision wedge biopsy is typically used to obtain a sample of skin that includes the dermis. If noncaseating granulomas are found, tissue culture may be necessary to exclude infectious causes. The pathologic differential diagnosis of sarcoidal granulomas includes cheilitis,

granulomatosa, cutaneous Crohn's disease, deep fungal infections, foreign body reactions, leishmaniasis, mycobacterium infections, papular acne rosacea, protozoa and late secondary and tertiary syphilis[21].

CONCLUSION

In light of this case report and brief review of literature, health providers should have a high index of suspicion of systemic sarcoidosis when they evaluate patients with granulomatous skin lesion and psoriasis with respiratory symptoms.

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