

Unusual Manifestation of Cutaneous Sarcoidosis: A Case Report of Morpheaform Sarcoidosis

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Abstract- Sarcoidosis is multi organ disease with cutaneous manifestation in 20%-35% patients. Cutaneous sarcoidosis has variable manifestations that make it difficult to diagnose. So clinical, histopathologic and laboratory evaluation is needed for diagnosis. Most of cutaneous lesions presents as nodul, maculopapule and plaque. Morpheaform lesion is a rare presentation of cutaneous sarcoidosis. This case had multiple indurated scaly plaques resemble morphea with granulomatous pattern in histopathologic examination. The patient responded to prednisolone in addition to hydroxychloroquine.

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Introduction

Sarcoidosis is a rare systemic, multi organ disease of unknown cause. The most common organ involved in sarcoidosis is lung, but cutaneous manifestations occur in 20%-35% of patients and can be as the initial presentation (1). Due to variety in the cutaneous manifestations, a classification is used to specify the lesions: specific and nonspecific. Specific lesions have non-caseating granulomas on histopathological view and in clinical pictures present as nodular, maculopapular, plaque type and infiltrative lesions. Nonspecific lesions have reactive pattern in histopathological examination and on clinical evaluation commonly presents as erythema nodosum in acute phase of the disease (1-3). Wide variety of manifestations, make the diagnosis of sarcoidosis challenging. Here we report an unusual manifestation of sarcoidosis that is similar to morphea in clinical examination but showed granulomas in histopathological assessment.

Case Report

A 17 year old boy came to dermatology clinic with an old erythematous scaly plaque on his face and 3 erythematous indurated plaques on his forearm, hand and foot with yellowish discharge. His problems has started since 9 years ago with an erythematous papule on

his right cheek. The papule was diagnosed as leishmaniasis and he took herbal drugs. Forever after a while the papule progressed to a large erythematous plaque with scaling. After 2 years patient developed same papule on his right forearm and received glucantime as treatment of leishmaniasis. But his disease progressed over the time. With diagnosis of morphea, the patient received methotrexate about 3 years ago which was not effective and patient developed new sclerotic lesion on his extremity. There was not any history of scar or trauma on the site of the plaque. On examination, a large indurated scaly plaque was seen on the right side of face (Figure 1). Also there were two indurated ulcerative plaques on his forearm and hand (Figures 2 and 3). The patient developed flexion contracture of the fingers due to plaque. At the time of examination some lesions had yellowish discharge.

Laboratory studies showed normal levels of serologic tests for vasculitis and autoimmune disease such as ANA, dsDNA Antibody (Ab), P-ANCA, C-ANCA, C₃, C₄, SM-Ab, SS-A Ab, SS-B Ab, Scl-70 Ab. Liver function test showed elevated levels of alanine aminotransferase (79 IU/L, 2-31 IU/L), alkaline phosphatase (1193 IU/L, 64-304 IU/L), GGT (402 IU/L, 5-40 IU/L). Microbiology culture of yellowish discharge showed heavy growth of pseudomonas after 3 days. His chest CT scan was normal without hilar lymphadenopathy or parenchymal change.

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Figure 1. Large indurated scaly plaque on the right side of face that progressed over years.



Figure 2. Indurated ulcerative plaques on his hand with yellowish discharge. This lesion made flexion contracture in fingers.



Figure 3. Indurated ulcerative plaques on his forearm.

Unusual manifestation of cutaneous sarcoidosis

Abdomen pelvic sonography showed minimally irregular liver borders which was in favor of parenchymal damage.

Skin biopsy of the forearm lesion showed chronic granulomatous inflammation with severe fibrosis and tiny foci of calcification. Fite stain for lepra bacillus and Acid fast stain for tubercle bacillus were negative. KOH prep was negative for fungi. More over no Leishman body was seen. PCR for tuberculosis and leishmania were negative. Liver biopsy was done due to elevated liver enzymes and abnormal sonography findings, which was indicative for early septal cirrhosis.

After all, the morpheaform of cutaneous sarcoidosis was diagnosed regarding to clinical and histopathological findings. So systemic therapy of corticosteroid (prednisolone, 50mg/d) in addition to hydroxychloroquine was started for him. Patient received antibiotic for his ulcer infection. After 2 weeks patient reported improvement of his lesions

Discussion

Sarcoidosis is diagnosed mainly by both clinical and histopathological findings. As sarcoidosis presents with variety of manifestations, diagnosis of this disease becomes difficult. So sarcoidosis is known as "great imitator" in dermatology (4). Cutaneous lesions are classified to specific and nonspecific based on presence of typical granulomas of sarcoidosis. In specific lesions histopathological assessment of skin biopsy shows non-caseating epithelioid cell granulomas, paravascular and perivascular inflammation while in nonspecific lesions non-diagnostic inflammatory reaction can be seen (2,5). Granulomatous pattern in skin biopsy had many differential diagnoses such as tuberculosis, fungal infection, leishmaniasis, rheumatoid nodules, Melkersson-Rosenthal syndrome and foreign body. So these pathogens should be excluded by staining to confirm the sarcoidosis (2). Therefore skin biopsy is not sufficient solely for diagnosis (6). The lesions of our patient were specific as inflammatory granulomas were diagnosed in skin biopsy. In clinical examinations, lesions had morphea characteristics which were correlated with severe fibrosis in biopsy.

Up to now just 6 cases of morphea form sarcoidosis were reported. Just 2 of them had cutaneous involvement without any systemic manifestations (7,8). Morphea is diagnosed by sclerosis of skin and associated with indurations in clinical examination. It has been showed that morphea is autoimmune disorder of skin (9).

Recent researches show that just 30% of cutaneous sarcoidosis lead to systemic sarcoidosis. Pulmonary involvement is reported in 90% of systemic sarcoidosis. These patients presents with cough, dyspnea, hemoptysis. In pulmonary involvement pulmonary fibrosis can be diagnosed in 10% to 15% of cases. In 50% to 80% of patients with systemic sarcoidosis the liver and spleen are involved. Hepatic sarcoidosis is 2 times common in African American. Fever, fatigue, weight loss and abdominal pain are characteristics of hepatic sarcoidosis although most of these cases are asymptomatic. If hepatic sarcoidosis is suspicious then liver function test, liver biopsy is indicated (10). In this patient alanine aminotransferase, alkaline phosphatase and GGT were elevated but early septal cirrhosis pattern of liver biopsy was not in favor of sarcoidosis. Elevated enzyme level and cirrhotic pattern of the liver can be attributed to methotrexate. Arthralgia, arthritis, muscle nodules can be diagnosed in systemic sarcoidosis which is indicative of musculoskeletal involvement (4). Our patient did not have any manifestation of pulmonary, hepatic and musculoskeletal involvement.

Most of the lesions in cutaneous sarcoidosis are refractory to treatment and recurrence is common after successful treatment. Topical steroids are used in localized lesions, although they do not penetrate the lesion adequately. Systemic treatments are used in multiple and progressive lesions. Systemic glucocorticoids are reported as most effective agent. But many patients do not respond to steroids well. Hydroxychloroquine, methotrexate, thalidomide can be effective in refractory patients (4).

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