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Case Report

Nearly common but missed confounder; Sarcoidosis!

Kausar Rehana, Anum Ilyas and Muhammad Mudassir Shafiq
Department of Pulmonology, Pakistan Institute of Medical Sciences, Islamabad

Abstract:
Sarcoidosis is a great masquerader. Its unusual presentations may be completely forgotten. We here describe here a case of sarcoidosis who presented with dermal manifestations of the disease.
Keywords: Sarcoidosis, Non caseating granuloma, Dermal sarcoidosis

Introduction

Pulmonary diseases have varying manifestation including symptoms from head to toe and patients with pulmonary disease can present first time in other domains. Sarcoidosis is a chronic granulomatous condition with unknown cause. Because patients are free of symptoms in most of the cases, sarcoidosis should be considered in differential diagnosis of non-caseating granuloma if noted in biopsies, performed for other reasons. Patients of sarcoidosis can present in any department including dermatology, neurology, ophthalmology and rheumatology. Here we present a similar case of a young male with multiple skin lesions.

Case summary:

A 33 year old male, with no pre-morbids presented in dermatology out door with skin lesions that had been here for the last 6 months, they were multiple, popular and nodular more on nose, right cheek and upper back but without any symptoms. (Figure1) There was no history of fever, joint pains, weight loss, eye or any other systemic complaints. Past and family history was not contributory. He underwent relevant examinatio n; among which skin biopsy showed non-caseating granulomas. (Figure2) He had intermittent complaint of dry cough for which he was sent to pulmonology department for further workup. His systemic examination was unremarkable. The chest x-ray showed bilateral hilar prominences and widening of right para-tracheal region. (Figure3) Thoracic CT scan with contrast was performed and was reported as bilateral axillary lymphadenopathy, significant lymph node sinpre-trachael, right para-trachael, carinal, subcarinal, prevascular, region and around aorta and inferior vena cava (Figure 4) (later confirmed on USG Abdomen).The findings reported principally favors diagnosis of sarcoidosis. Patient's skin biopsy was re-evaluated by the pathology department and was confirmed to be non-caseating granulomas. Findings were consistent with sarcoidosis. Increased levels of ACE were found (161U/L; normal range 8-65U/L). Rest of his blood chemistry and ESR were normal. After treatment with prednisolone 1.5mg/kg/day, he had near-complete resolution of his skin lesions (Figure1) at the end of six months and his ACE levels were reduced to normal.

Author for Correspondence:
Dr. Abdul Khalid
Associate professor medicine AJK-Medical College Muzaffarabad Azad Kashmir
e-mail: khalid_awanajk@yahoo.com

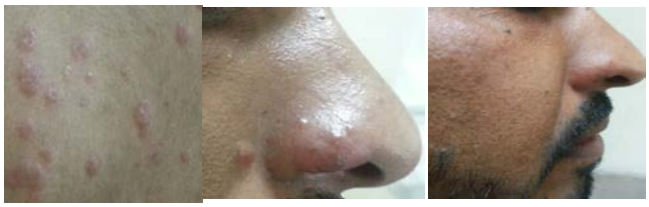

Figure 1: Multiple popular and nodular lesions on upper back, cheek and nose before and after treatment

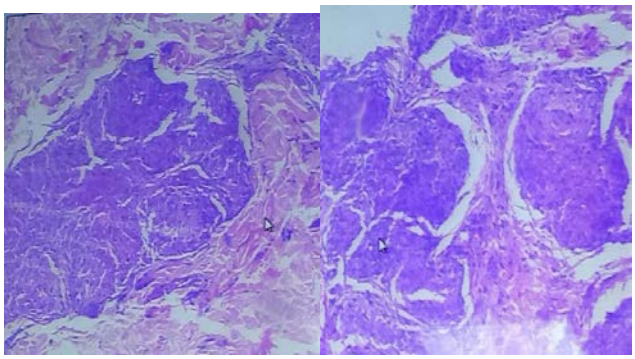

Figure 2: Histopathological section showed non-caseating granulomas without prominent lymphocytosis.

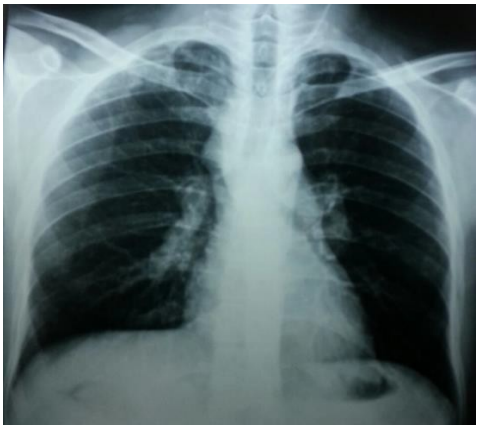

Figure 3: CXR showing bilateral hilar prominence and widening of right paratrachael region.

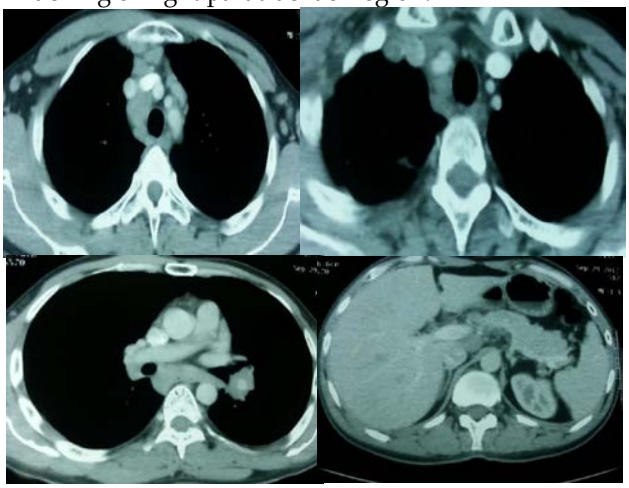

Figure4: CT Scan Chest showing lymphadenopathy in bilateral axillary, pretrachael, right paratrachael, precarinal, subcarinal, prevascular, bilateral hilar region, around the aorta and inferior vena cava

Sarcoidosis is a systemic granulomatous disease of multisystem affecting any organ of body; lungs and lymphoid system are the most commonly affecting organs with frequency of 90% and 30% respectively. (1) Extra pulmonary involvement is reported in 30% of patients, and the abdomen is the most common extra pulmonary site with a frequency of 50%–70%. Although intra-abdominal sarcoidosis is usually asymptomatic, its presence may affect the prognosis and treatment options. The lesions are less characteristic and may mimick neoplastic or infectious diseases such as lymphoma, diffuse metastasis, and granulomatous inflammation.

Skin lesions may appear at any stage of the disease and specific skin lesions may occur in 9% to 37% of patients. (2,3) However, cutaneous lesions are the sole manifestation of sarcoidosis in approximately 10% of patients. (2) Lesions often appear at onset of systemic illness providing a valueable opportunity for early diagnosis (4,5) as was in our case. Cutaneous sarcoidosis may

Develop with or without systemic involvement. Sarcoidosis becomes a diagnostic challenge as skin lesions present with wide spectrum of different morphologies especially with no systemic involvement. (6,7)

Cutaneous sarcoidosis was initial manifestation in approximately one third of our patients and was compatible finding with literature data. (2,3) A recognition of cutaneous lesions is important because they provide a visible clue to diagnosis especially if they have a specific appearance. These lesions also are an easily accessible source of tissue for diagnosis, as was in our case so whenever a patient presents with skin lesions; skin biopsy either punch or excisional biopsy should be done in order to establish a proper diagnosis.

CT scan chest with contrast was thought to be an accurate and effective examination for sarcoidosis and mediastinal diseases. The typical manifestation included bilateral hilar lymphadenopathy symmetrically accompanied with mediastinal lymphadenopathy near both the pulmonary hila, which could be presented in about 80%–90% of these patients. (5) The intrapulmonary lesions included alveolar nodules, granulomatous nodules, pulmonary fibrosis and soon. (8) We were able to diagnose a patient of sarcoidosis on skin biopsy and found to have abdominal lymphadenopathy on CT scan, enlarge lymph nodes are detected in only 30% of patients with particularly in the porta hepatis, para-aortic region, and the celiac axis. Unlike lymphoma, the lymph nodes are typically smaller than 2 cm in diameter and more discrete rather than confluent. Compared to lymphoma, involvement of the retrocrural area is less common.

Conclusion

In conclusion, patients with sarcoidosis can present with skin lesions having non-caseating granulomas on skin biopsy along with an elevated serum ACE level are associated with progressive disease. Skin biopsy is a simple and accurate technique that can be easily performed for the diagnosis of sarcoidosis and also simple measures should be taken for diagnosis as in our case patient also had abdominal lymphadenopathy but skin biopsy proved sarcoidosis. The case facilitated imaging diagnosis of the sarcoidosis,

which would aid accurate diagnosis by combining with biopsy.

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CONTRIBUTION OF AUTHORS	
Author	**Contribution**
Kausar Rehana	A,B,C,D,E,F
Muhammad Mudassir	A,B,C,D,

KEY FOR CONTRIBUTION OF AUTHORS:

A. Conception/Study Designing/Planning
B. Experimentation/Study Conduction
C. Analysis/Interpretation/Discussion
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