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# Sarcoidosis: A Great Masquerader-Case Report

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**ABSTRACT**
*Sarcoidosis is systemic disorder of unknown origin characterized by granulomatous inflammation in different organs , most frequently in lungs and affects people of all racial and ethnic groups. Here we present a case of sarcoidosis with respiratory symptoms and an unusual site of subcutaneous nodule. A high index of clinical suspicion led to the correct diagnosis in this atypical presentation.*

## INTRODUCTION

Sarcoidosis is a multiorgan disorder; patients may present to different specialties. The clinical presentations vary depending upon the ethnicity, duration of illness, site and extent of organ involvement and activity of granulomatous process. It is an unusual cause of fever of unknown origin. It has been suggested that sarcoidosis is a consequence of genetically susceptible individuals exposed to specific environmental agents. Acute presentations are Lofgren's syndrome (b/l hilar lympahdenopathy, ankle arthritis, erythema nodosum and constitutional symptoms) or Heerfordt's syndrome (fever, parotid gland enlargement, facial plasy and uveitis). Chronic sarcoidosis is insidious in onset and its symptoms are related to the organs involved (lungs, eye, heart, skin, lymph nodes, etc).

## CASE REPORT

A 30 year old house wife with no significant past history or family history presented with 3 months history of dry cough; without dyspnea, chest pain or hemoptysis.

On examination her vitals were normal. General physical examination was normal except for a non tender subcutaneous nodule on the right sole of foot, about 2x2 cm in size.

Systemic examination including respiratory system and ophthalmologic examination were found to be normal.

Investigations revealed normal routine blood examination except for an ESR of 90mmHg. LFT, RFT, lipid profile, TFT, blood sugar, serum electrolytes, urine routine were normal. Viral markers-negative.

Blood Culture including fungal culture-negative. Serum polysaccharide Antigen Assay test for Histoplasma-Negative.

Ophthalmologic examination ruled out any evidence of eye involvement.

Echo/ECG/USG abdomen- all normal.

X ray chest- bilateral hilar lymphadenopathy without any parenchymal lesions

HRCT-chest: bilateral hilar and mediastinal lymphadenopathy with perifissural nodules in both upper lobes

In view of respiratory symptoms with bilateral hilar lymphadenopathy and high ESR, excision biopsy of the subcutaneous nodules was done. The histopathological report was suggestive of Sarcoidosis. Cardiac MRI- normal.

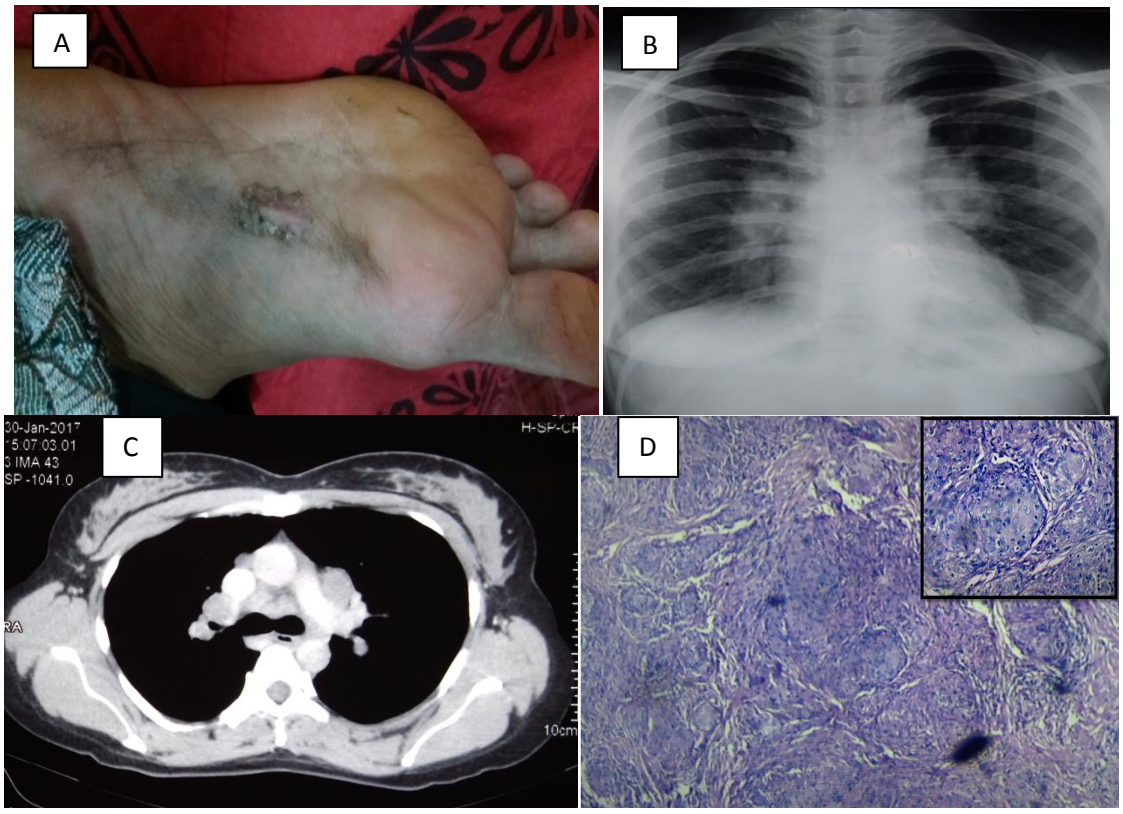


**Fig.1** A, Skin lesion over sole of right foot. B, X ray chest- bilateral hilar lymphadenopathy. C, HRCT-chest showing bilateral hilar and mediastinal lymphadenopathy. D, Biopsy: H and E low power showing noncaseating granuloma; (inset) high power showing epithelioid granuloma consisting of giant cell, epithelioid cell and sparse lymphocytes.

## DISCUSSION

The clinical spectrum in sarcoidosis is protean , ranging from an abnormal chest radiograph in an asymptomatic individual (upto 50% patients) to severe multiorgan involvement. Sarcoidosis skin lesion mimics various common dermatologic conditions, causing great confusion for the diagnosis and posing problems for management. Awareness of these varied morphologic presentations is essential for the early diagnosis and management of the master mimicker - cutaneous sarcoidosis. Specific skin lesions include skin plaques, maculopapular eruptions, subcutaneous nodular lesions, erythroderma, ulcerations, scarring alopecia, infiltration of old scar and lupus pernio. Lupus pernio is the most specific skin lesion of sarcoidosis. it is an indolent, red purple or violaceous disfiguring skin lesion that usually affect nose, cheeks, ears and forehead. It is associated with a chronic fibrotic pulmonary sarcoidosis. Non specific skin lesions include erythema nodosum (13-20%), usually suggests a good prognosis. This lesion is not diagnostic of sarcoidosis, as it can be seen with infections, vasculitidis, drug reactions and neoplasms. Less common non specific lesions

include acquired icthyosis, calcinosis cutis, erythema multiforme and nail clubbing.

Pulmonary involvement is the most common visceral manifestation occurring in 95% patients ranging from hilar lymphadenopathy (50%) to ILD with alveolitis, pleural effusions are rare with less than 5%. Sarcoid granuloma develop throughout the lung parenchyma , but they present mostly prominently in the upper 2/3$^{rd}$ of lung. They are common along the bronchovascular bundles, subplureal locations, intralobular septae and also in the airways. Symptoms include dry cough (30%), dyspnea (28%), chest pain (15%). wheezing can occur when there is an endotracheal involvement. Unlike most ILD that demonstrate a restrictive ventilatory defect, pulmonary sarcoidosis may manifest as obstructive lung disease or a restrictive lung disease or obstruction and restriction may occur concomitantly. Gas exchange remains normal until late in the disease course. Resting hypoxia and reduction in DLCO are not commonly seen except with fibrocystic (stage 4) or pulmonary hypertension which may develop.

Arthritis (4-38%) and arthralgia are seen. Lofgren's syndrome is a triad of acute arthritis, erythema nodosum and bilateral hilar lymphadenopathy. There is excellent response to corticosteroid therapy. Acute histoplasmosis may resemble lofgren's syndrome and must be excluded by serology and culture.

Ocular involvement (25%) is typically bilateral and can be acute anterior uveitis which is the most common manifestation or chronic anterior. Hepatomegaly (20%), splenomegaly (10%), heart involvement (5%) may occur. Hypothalamic pituitary axis may be involved and classically present as diabetic insipidus. Kidney and GIT organs are rarely affected. Vasculitis of any size vessel has been described.

Tissue biopsy is the gold standard for confirming a clinicoradiograph diagnosis of sarcoidosis. Well circumscribed, non caseating granulomas of epitheloid type are widely distributed and have been reported in many organs. They are most commonly found in the lung (90% if abnormal chest X ray,40% if normal chest X ray, 83% using endobronchial USG guided transbronchial FNAC of intrathoracic lymph nodes), liver and muscles (50-80%), minor salivary glands (36%) and bone marrow (17%). When the following organs are clinically abnormal, granuloma can be demonstrated in skin (90%), parotid gand (90%), lymph nodes (90%), synovium (80%) and heart (20%). Granuloma may produce elevated ACE levels in blood and may significantly enhance the production of 1, 25-dihydrocholecaliferol which is responsible for clinical hyeprcalcemia and hypercalciuria. ACE is produced by epithelial cells and macrophages at the periphery of granuloma in response to an ACE-inducing factor released by T cells. ACE is also elevated in military TB, histoplasmosis, silicosis, asbestosis, Kaposi sarcoma with HIV infection, hyperthyroidism, diabetes, cirrhosis, Gaucher's disease and hypersensitive pneumonitis. Additionally ACE levels are influenced by ACE gene polymorphism (DD allele). Therefore an elevated ACE level may be supportive, especially if more than twice the upper level of normal, but not diagnostic of sarcoidosis.

This patient was started with low dose oral prednisolone and hyroxychloroquine and now shows improvement.

## CONCLUSIONS

The clinical spectrum of sarcoidosis varies. Skin lesions of sarcoidosis are like the tip of an iceberg indicating more changes in other organs .Atypical presentations pose diagnostic challenge in sarcoidosis. In our case of a non tender, subcutaneous nodule in an unusual site in a patient with only respiratory symptoms triggered a high index of clinical suspicion which led to the correct diagnosis of sarcoidosis. This case underlines the importance of proper clinical examination and need to identify accessible and less invasive sites for tissue biopsy to reach correct diagnosis.