

## Case Report

# Cutaneous sarcoidosis with protean manifestations: a rare case report

Avinash Pravin, Shreya Srinivasan, Jayakar Thomas\*

Department of Dermatology, Sree Balaji Medical College Hospital, Chromepet, Chennai, Tamil Nadu, India

**Received:** 12 February 2019

**Accepted:** 22 March 2019

**\*Correspondence:**

Dr. Jayakar Thomas,

E-mail: [jayakarthomas@gmail.com](mailto:jayakarthomas@gmail.com)

**Copyright:** © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

### ABSTRACT

Sarcoidosis is a multisystem granulomatous disease of unknown origin. The lung is the most commonly affected organ, but skin is frequently involved. Skin involvement is important because it may be the presenting sign of systemic sarcoidosis. Here, we report a rare case of sarcoidosis with systemic manifestations, in a 39 year old male patient.

**Keywords:** Sarcoidosis, Lung involvement, Imitator

## INTRODUCTION

Sarcoidosis was described initially by Sir Jonathan Hutchinson in 1875 and cutaneous sarcoidosis (Lupus pernio) by Besnier in 1889. The term Sarcoidosis is derived from Caesar Boeck's 1899 report of what he described as 'multiple benign sarcoid of the skin' because he believed the lesions resembled sarcomas but were benign.<sup>1</sup> It is a systemic granulomatous disorder of unknown origin that commonly involves the lungs.<sup>2</sup> Cutaneous manifestations of sarcoidosis are seen in only upto one-third of patients and they may be the first clinical sign of the disease.<sup>2</sup> The common sites of cutaneous sarcoidosis are the head and neck region. Skin lesions can be papular, nodular, plaque, patches forms, out of which the papular form is the commonest. Variants of sarcoidosis include lupus pernio, subcutaneous, ichthyosiform, angiolutoid and ulcerative. Sarcoidosis occurs more frequently between 20 and 40 years of age and there is a second peak of incidence around age of 60. It is more common in women than men, less common in elderly and rare in children. Here we report a rare case of sarcoidosis with protean manifestations.

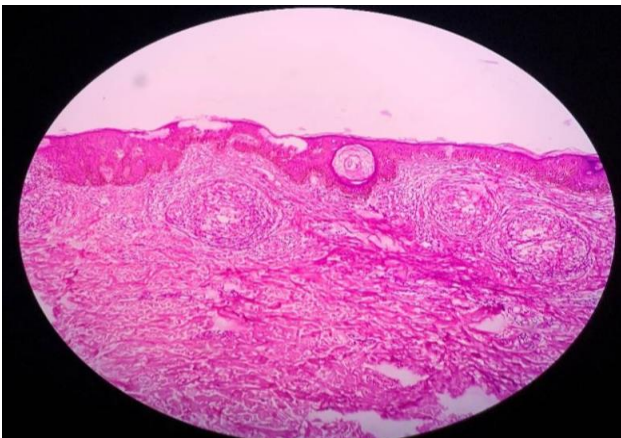
## CASE REPORT

A 39-year-old male patient presented to the skin OPD with complaints of multiple skin coloured raised lesions on the face, neck, axillae and groin for the past 2 years. Patient was apparently normal 3 years back after which he noticed dark coloured flat lesions on bilateral legs and then later started developing the aforementioned lesions. He has a positive history of intermittent productive cough for the past 3 years. He reports a left sided facial palsy one year back. On examination, multiple skin coloured to pigmented papules were seen over the face, neck, bilateral axillae and groin. Pigmentation could be appreciated over the legs. Chest X-ray showed few fibrotic strands in the lateral aspect of left middle and upper zone as well as in the right middle zone (Figure 3). HRCT chest revealed centrilobular nodules and bilateral hilar lymphadenopathy. Pulmonary function test was suggestive of mild restriction. Pulmonologist's opinion was obtained and the patient was deemed to be pulmonary sarcoid stage two. Serum calcium levels were normal but serum angiotensin converting enzyme levels were elevated. Histopathology revealed well circumscribed collections of epithelioid cells and few lymphocytes in the dermis with epidermal atrophy.

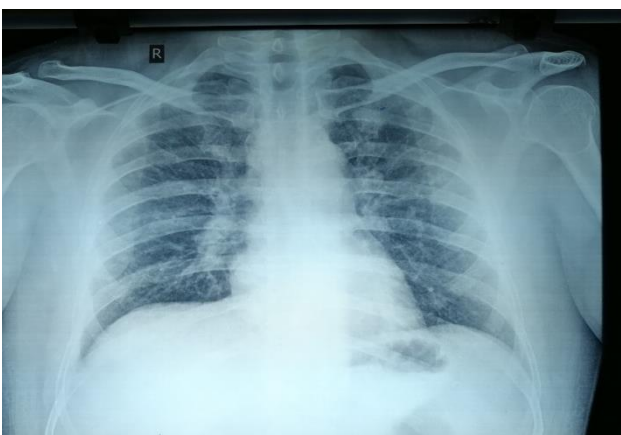
Based on the above said findings, we made a diagnosis of cutaneous sarcoidosis with protean manifestations.



**Figure1: Clinical photograph showing multiple papules over the neck.**



**Figure 2: Photomicrograph of the biopsy specimen under low power H&E stain shows well circumscribed aggregates of histiocytic epithelioid cells with few lymphocytes.**



**Figure 3: Chest X-ray showed few fibrotic strands in the lateral aspect of left middle and upper zone as well as in the right middle zone.**

## DISCUSSION

It is a multisystem granulomatous disorder of unknown origin that mainly involves the lungs, mediastinum, peripheral lymph nodes, eyes and skin. There is an increased risk of developing lympho-proliferative disease, mainly Hodgkin lymphoma. Several case reports have described the concomitant association of sarcoidosis with number of autoimmune diseases particularly Graves disease, Hashimoto's thyroiditis. Syndromes associated with Sarcoidosis include Lofgren's syndrome characterized by erythema nodosum, hilar adenopathy, fever, arthritis and Heerfordt's syndrome characterized by parotid gland enlargement, uveitis, fever, cranial nerve palsy.

Cutaneous manifestations are usually asymptomatic and mimic a number of conditions. The most common cutaneous presentation is the papular form, which often has a translucent yellow brown appearance appreciated better in Caucasians. The yellow brown colour is likened to an 'apple jelly' appearance on diascopy. Specific lesions like lupus pernio and erythema nodosum are associated with severe systemic involvement and acute benign disease respectively<sup>3</sup>.

The lung is the most commonly involved organ. Patients can present with dyspnea, cough, chest pain and wheezing. Chest X-ray is abnormal in more than 90% cases showing bilateral hilar lymphadenopathy. Abnormal pulmonary function test also reported in 50-70% of cases and is usually indicative of restrictive pulmonary disease.

Eye involvement with sarcoidosis is potentially vision threatening and can be the first manifestation of the disease.<sup>4</sup> Uveitis is the most common ocular manifestation and can lead to cataracts and glaucoma. Other ocular manifestations include conjunctivitis, lacrimal gland involvement causing kerato-conjunctivitis sicca & optic neuritis which may rapidly lead to vision loss.

Hepatic sarcoidosis is present in more than one-half of patients but signs of organ dysfunction due to hepatic sarcoidosis is less common. Cardiac involvement is found only in 5% of patients in the form of arrhythmias, conduction block and sudden death. Any portion of the CNS or peripheral nervous system may be affected by sarcoidosis. The Facial nerve is the cranial nerve most commonly involved.<sup>5</sup> In fact, it is common for Bell's palsy to be the first manifestation of sarcoidosis. The sinuses and upper airway are commonly involved with sarcoidosis in a condition known as SURT: (sarcoidosis of upper respiratory tract). Nasal SURT is often associated with lupus pernio. Sarcoidosis can cause disorder of calcium metabolism that results in hypercalcemia. Histopathology of sarcoidosis shows well-circumscribed non-caecating granulomas with

aggregates of epithelioid histiocytes surrounded by few to no lymphocytes giving the term, naked granuloma.

Sarcoidosis is a great imitator of a number of dermatological problems like leprosy, tuberculosis, lymphoma cutis to name a few and is a basically a diagnosis of exclusion.

Sarcoidosis is usually refractory to treatment and involves topical or intralesional steroids for localized involvement and systemic glucocorticoids for systemic involvement. Other modalities of treatment include antimalarials, methotrexate, tetracyclines, thalidomide and infliximab.

## CONCLUSION

In India, the incidence of sarcoidosis is uncommon and is usually a diagnosis of exclusion. When a patient is suspected to have cutaneous sarcoidosis, a systematic examination should be done to rule out internal involvement. This case has been reported because of the rarity of cutaneous sarcoidosis with protean manifestations.

*Funding: No funding sources*

*Conflict of interest: None declared*

*Ethical approval: Not required*

## REFERENCES

1. Boeck C. Multiple benign sarcoid of the skin. J Cut Genito-Urin Dis. 1899;17:543-50.
2. Travis WD, King TE, Bateman ED, Lynch DA, Capron F, Center D, et al. American Thoracic Society/European Respiratory Society international multidisciplinary consensus classification of the idiopathic interstitial pneumonias. Am J Respiratory Critical Care Med. 2002;165(2):277-304
3. Chadha DS, Vasdev V, Gupta S. Sarcoidosis masquerading as a dermatological problem presenting a diagnostic challenge in an aviator-a case report. Ind J Aerospace Med. 2009;53(1):52.
4. Ohara K, Judson MA, Baughman RP. Clinical aspects of ocular sarcoidosis. European Respiratory Monograph. 2005 Sep 1;32:188.
5. Stern BJ, Krumholz A, Johns C, Scott P, Nissim J. Sarcoidosis and its neurological manifestations. Arch Neurol. 1985;42(9):909-17.

**Cite this article as:** Pravin A, Srinivasan S, Thomas J. Cutaneous sarcoidosis with protean manifestations: a rare case report. Int J Res Dermatol 2019;5:437-9.