

## **Sarcoidosis masquerading as a dermatological problem presenting a diagnostic challenge in an aviator – A case report**

Wg Cdr DS Chadha\*, Wg Cdr V Vasdev<sup>+</sup>, Wg Cdr S Gupta<sup>#</sup>

### **ABSTRACT**

A 36 year old IAF pilot presented with a two years history of recurrent nodules and plaques on his back, shoulder, and face. Skin biopsy showed diffuse non-necrotizing granulomatous lesion infiltrated with numerous multinucleated giant cell and lymphocytes. Missed diagnosis of cutaneous tuberculosis and later leprosy was made for which he received antituberculous and antilepromatous drugs. A repeat skin biopsy upon recurrence of skin lesion in the presence of bilateral hilar lymphadenopathy led to the diagnosis of sarcoidosis. He was treated with oral steroids with which his lesions regressed. Diagnostic challenges faced in a case of cutaneous sarcoidosis and its associated aeromedical concerns will be discussed.

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**Key words:** Sarcoidosis, lymphadenopathy, giantcells.

### **Introduction**

Sarcoidosis is a rare chronic granulomatous disease of unknown etiology that primarily affects individuals between 30 and 50 years of age. It is usually characterized by the presence of non-caseating, granulomatous, epithelioid tissue at the sites affected with marked variability in prevalence and pattern of organ involvement [1]. The diagnosis is based on the consistent clinical and radiological findings, supported by histological picture. In approximately one-half of cases, the disease is detected incidentally by radiographic abnormalities noted on a routine chest x-ray prior to the development of symptoms.

Approximately 20 to 35% of patients with sarcoidosis have cutaneous involvement. Although cutaneous lesions may present at any time during the course of the disease, they often present early after disease onset. Cutaneous sarcoidosis, reported rarely from India, is known as one of the “great imitators” in dermatology as lesions may assume a vast array of morphologies, masquerading as a wide range of disorders from benign appendageal growths to malignant Kaposi

sarcoma [1-6]. Therefore, delayed or missed diagnosis often occurs because it may resemble leprosy, tuberculosis, lymphoma cutis [pseudolymphoma] and other granulomatous diseases. We present here a case of cutaneous sarcoidosis which presented as a diagnostic problem.

### **Case Report**

A 36 yrs old airforce transport pilot presented for the first time in 1999 with maculopapular skin lesions involving face, shoulders and back. He was initially seen at a civil hospital where he was given a presumptive diagnosis of cutaneous tuberculosis. The detailed evaluation and reports were not available for perusal. He received a full course of ATT along with steroids with which his skin lesions resolved. However in 2003 he had recurrence of skin lesions and was diagnosed as a case of leprosy

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\* Cl Spl (Med&Cardiology) Command Hospital, Bangalore

+ Cl Spl (Med), AH (R&R), New Delhi

# Cl Spl (Med&Neurology), Command Hospital, Bangalore



**Figure 1: Maculopapular skin lesions and plaques over back**

based on histopathological report of non caseating granuloma and PCR positivity for *M leprae*. Acid fast bacilli were not demonstrated in the microscopic examination of the specimen. Other routine investigations including the mantoux test were normal. He had exacerbation of skin lesions while on treatment and it was attributed to Type-I lepra reaction. Steroids were exhibited that resulted in partial remission of lesions. While on anti leprosy treatment for almost a year he was reviewed at aeromedical evaluation center and was found to have maculopapular skin lesions involving face, arms and thoracic region (Figure 1).

Systemic examination was unremarkable. Chest radiograph done at this stage revealed bilateral hilar adenopathy without any pulmonary parenchymal lesions (Figure 2).

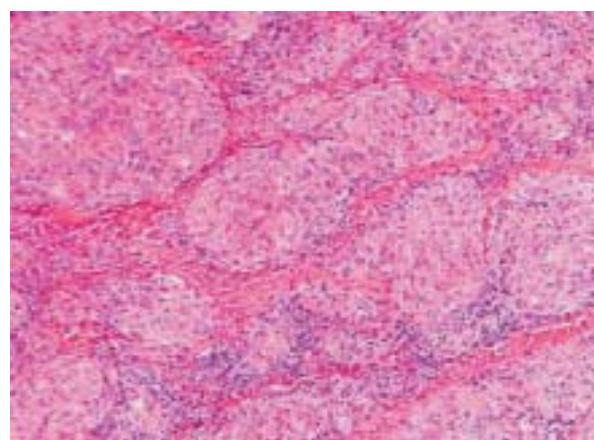
CT scan chest showed few enlarged lymph nodes in right paratracheal and precarinal region.



**Figure 2: Hilar lymphadenopathy in chest radiograph**

PFT (spirometry) and diffusion studies were within normal limits. The diagnosis of sarcoidosis was considered and subsequently confirmed on histopathological examination (Figure 3).

Further detailed work up (angiotensin converting enzyme levels, serum calcium, 24 hours urinary calcium, pulmonary function testing, resting 12 lead Electrocardiogram, Echocardiogram, 24 hours Holter monitor, CT Scan Brain & slit lamp examination) showed no evidence of ophthalmologic, neurologic, cardiovascular or renal involvement. He showed good response to treatment with steroids. Follow up over 6 months showed complete regression of skin lesions and hilar



**Figure 3: Skin biopsy showing sarcoid granulomas**

lymphadenopathy, he awaits waiver for flying after further close observation.

## **Discussion**

Cutaneous sarcoidosis may occur at any stage of the disease but most often it is present at the onset of disease [2]. Recognition of cutaneous lesions is important because they provide a visible clue to the diagnosis and are an easily accessible source of tissue for histologic examination. However cutaneous lesions may masquerade as a wide range of disorders (leprosy, tuberculosis, lymphoma cutis, lupus vulgaris, leishmania etc) leading to missed and delayed diagnosis as was noted in the present case [3,4].

Patients with cutaneous sarcoidosis are usually asymptomatic. Skin lesions are polymorphous and are classified into three basic groups, erythema nodosum, scar sarcoidosis and skin sarcoid [4]. Skin sarcoid is characterized by specific cutaneous lesions which may take nodular, plaque, angiolupoid, or subcutaneous forms. Involvement may be mild or severe, and course may be self-limited or chronic [4,5]. Specific skin lesions like lupus pernio is associated with more severe systemic involvement, while erythema nodosum often indicates acute benign disease [2].

Cutaneous sarcoidosis is a great imitator and in patients with disease confined to skin establishing diagnosis may be challenging, even more so in developing countries where its prevalence is much lower than systemic tuberculosis and/or leprosy [5,6]. The current case continued to occupy the physician's attention and was erroneously administered ATT and antilepromatous drugs with no favorable response. Antilepromatous therapy was given based on the histopathological picture and PCR positivity for *M leprae*, this highlights the

lack of specificity of these in deciding therapy. However it was bilateral hilar lymphadenopathy noted on chest radiograph which raised the suspicion and later confirmed the diagnosis of sarcoidosis. Shegal et al have noted similar finding in their case [8]. Sarcoidosis confined to skin is very rare and it is invariably associated with some form of systemic component. Roughly 10% of sarcoidosis cases may develop systemic complications in the form of myocardial involvement (arrhythmias, conduction block, sudden death), restrictive pulmonary disease, CNS disease (cranial nerve palsies, encephalopathy, seizures), ocular complications (uveitis, iritis, chorioretinitis) and renal calculi [1]. There might be marked variability in systemic affection; hence it is prudent to evaluate all patients in detail.

The treatment of cutaneous sarcoidosis is often frustrating because lesions may be refractory to treatment or may recur following successful treatment. For localized involvement, topical or intralesional steroids may be used. Systemic glucocorticoids are reserved for progressive pulmonary disease, cardiac involvement, CNS disease, uveitis or hypercalcemia. Refractory cases may require therapy with hydroxychloroquine, methotrexate or thalidomide although randomized controlled trials with these agents are lacking [1-3].

Aircrew identified as having probable sarcoidosis should be grounded for a minimum of three months, to confirm the diagnosis histologically and determine the extent of disease [8]. Of primary interest to aviation medicine is the risk of cardiac or neurosarcoidosis (noted in about 5% of patients) which lead to permanent disqualification from flying. Those with active disease and on therapy with steroids should be restricted from flying. Medical certification is given to those who are off steroids

and have no evidence of systemic illness [9].

Present case highlights as to how cutaneous sarcoidosis, an easily treatable disorder, can masquerade other dermatological conditions resulting in delayed diagnosis with loss of highly skilled man power.

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