SARCOIDOSIS: Case report

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ABSTRACT:
A male patient presented with wide spread multiple cutaneous nodules affecting forehead, nostrils, upper arms, thighs and trunk. The nodules were asymptomatic and had a violaceous color and erupted over a period of 2 years. The clinical diagnosis of sarcoidosis was confirmed by skin biopsy. Despite the wide spread lesions no systemic involvement was detected. Patient responded to systemic steroid and is periodically checked for possible future systemic involvement

CASE REPORT:
A 34 years old male patient came to the dermatology clinic because of multiple asymptomatic cutaneous nodules affecting forehead, both nostrils, upper arms, thighs and trunk (Fig 1). The lesions began to appear 2 years ago in June 1997, 2 months after returning from Mecca after pilgrimage. The patient also lived in India for four years (1982 - 1986). The nodules were violaceous in colour, subcutaneous, raised and fixed to the skin, and most of them had a diameter of 10-12 mm. The forehead and nostril lesions were papular. The patient had no lymph gland, liver or spleen enlargement. Sensatious, lacrimal and salivary glands were normal. Investigations done were normal except for a raised ESR. The investigations included complete blood count, blood biochemistry profile, serum protein electrophoresis, compliment estimation, 24 hour urine calcium, serology for hepatitis and HIV. Tuberculin test was negative.
Fundoscopy and slit lamp examination for the eyes were normal. ENT examination showed normal upper respiratory tract. X ray chest showed slight prominance of right hilum and normal heart and lungs. The diagnosis was sarcoidosis and biopsy of one nodule showed sarcoid granuloma (Fig 2,3). Body gallium scan was requested. The diagnosis of Sarcoidosis has been confirmed and patient was put on systemic steroid 1 mg/kg daily and began to improve. No new lesions, some lesions cleared and some got smaller. The patient is followed up for evidence of systemic Sarcoidosis that may develop.

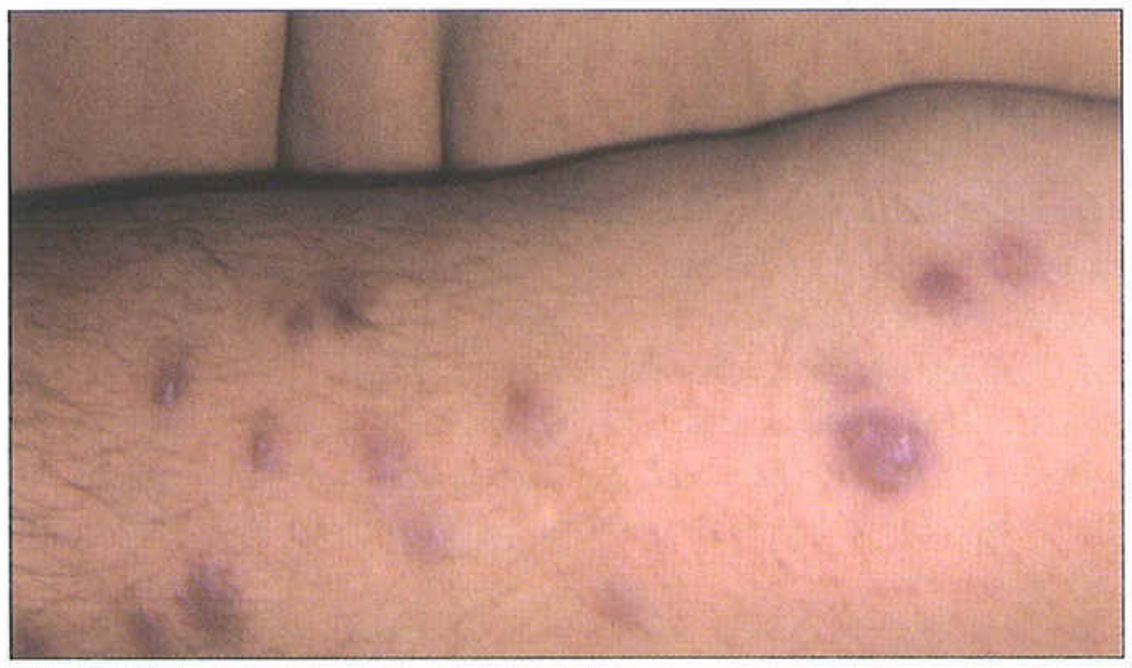

Fig. 1: Subcutaneous sarcoid nodules

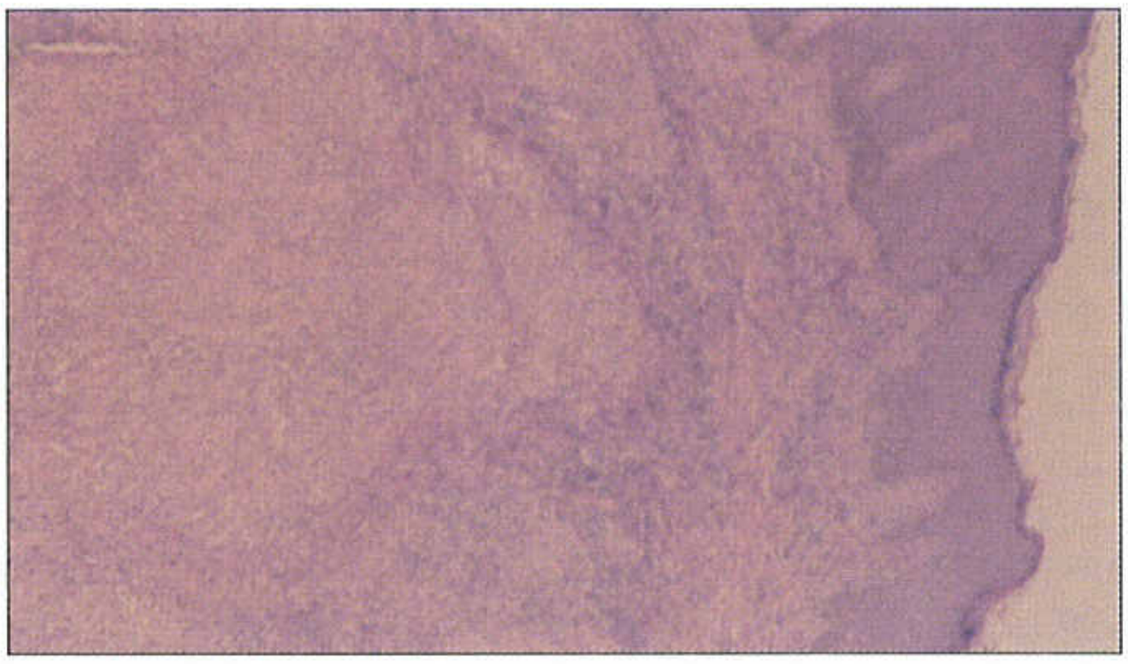

Fig. 2: Histopathology showing non-caseating sarcoid granuloma

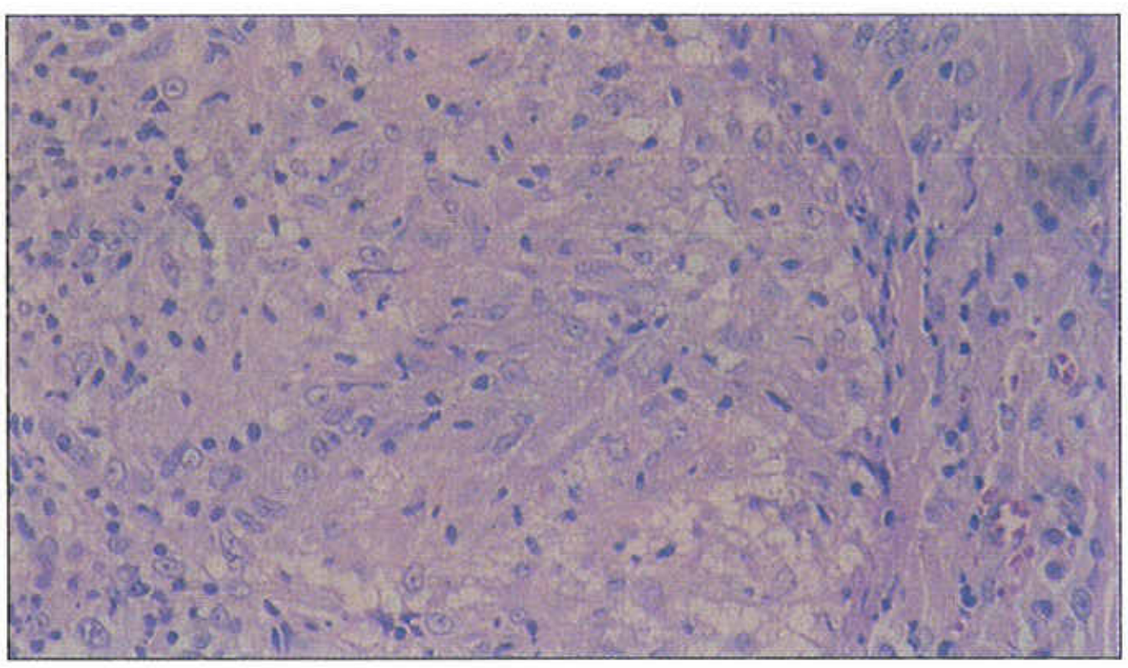

Fig. 3: Histopathology showing non-caseating sarcoid granuloma

DISCUSSION:
The aetiology of Sarcoidosis is unknown. The disease is characterized by formation of noncaseating granuloma which may involve any organ of the body. The main organs affected are lungs, mediastinal and peripheral lymph nodes, skin, eyes, parotid glands, liver, spleen and less frequently central nervous system, heart, upper respiratory tract and bones

(1,2). Histologic confirmation of the diagnosis of Sarcoidosis is necessary in most cases. The sarcoid granuloma shows a center of epithelioid cells surrounded by lymphocytes, macrophages,fibroblasts and occasional Langhans cells. Three types of inclusion bodies were described within giant cells viz the Schaumann body, the Asteroid body and the
residual body. The Kveim-Siltzbach test is reported to be reliable in confirming the diagnosis of Sarcoidosis in upto 90% of patients with active disease with only a 2% incidence of false positive result (3). Since the cause of Sarcoidosis remains unclear it is best regarded as a reaction pattern to an infectious agent or allergen (3,4).

Mycobacterium tuberculosis have been considered a causative agent for Sarcoidosis (5,6) Recent studies have detected RNA and DNA of M. tuberculosis in Sarcoidosis (7,8) Sarcoidosis is considered an antigen T-cell mediated immune response (9) In the affected organs activated T-cells and macrophages release a number of cytokines that stimulate granuloma formation (1) Most of these lymphacytes are T-helper cells (10) The large number of T-helper cells in affected sites may secondarily decrease their number in peripheral circulation resulting in a relative increase of suppressor cells and the occurrence of anergic state. (11)

The skin lesions in Sarcoidosis are classified as specific showing the characteristic granuloma and nonspecific lesion mainly erythema nodosum. The specific lesions are seen in 9 - 37% of patients (12) and include lupus pernio, infiltrated plaques, maculopapular eruption, subcutaneous nodules and infiltrations of old scars. Different types of skin lesions may occur in the same patient. The most common sarcoidal skin lesions are maculopapules, 13, infiltration of old scars from surgery, trauma, or acne.Venipuncture may become red purple and indurated and tend to persist according to the activity of Sarcoidosis (14) . Skin plaques appear as round or oval brownish red infiltrated patches most commonly located on limbs, face, scalp, back and buttocks (15)

The most typical specific skin sarcoidal lesion is lupus pernio which is a red purple violaceous indurated skin lesion which usually affects the nose, cheeks, ears, lips, forehead and may co-exist with Sarcoidosis of the upper respiratory tract and may be associated with pulmonary fibrosis, chronic uveitis and bone cysts. Sarcoidal skin lesions are the most often presenting sign of the disease (16) Periodical screening of patients with cutaneous lesions for systemic involvement should be done (1)

Three out of four patients with nasal rim skin lesion when screened for systemic involvement showed sarcoidosis of upper respiratory tract. (17) The most important nonspecific skin lesion in Sarcoidosis is erythema nodosum and its association with hilar lymphadenopathy is known as Lofgren syndrome which is the hallmark of acute and benign sarcoidosis (1). Clinically atypical granulomatous cutaneous Sarcoidosis includes extensive ulcerative lesions, psoriasiform plaques, hypopigmentation in black persons, verrucous and papillomatous lesions, icthyosiform changes, pustular folliculitis, papules in light exposed areas, lichenoid eruption, erythrodermic eruption, cicatricial alopecia, lupuss erythematosus like eruption, mutilating lesions, erythemas and plaques involving palms and soles (1)

TREATMENT:

Systemic steroids are used in treating Sarcoidosis. Antimalarial drugs and methatrexate are also tried. Topical or intralesional steroids and surgical excision of skin lesions was successfully tried. (18)

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