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Case report

CUTANEOUS SARCOIDOSIS: A RARE CASE REPORT

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ABSTRACT

Sarcoidosis is a Greek word (Sarco means flesh and Eido means type or like). Cutaneous sarcoidosis occurs in up to one third of patients with systemic sarcoidosis. This disease is characterised by the presence of non - caseating epitheloid cell granulomas in the skin. Cutaneous sarcoidosis presents as a diagnostic challenge to the dermatopathologists due to its varied presentations and almost identical histologic pictures. Hence, exclusion of infectious causes and compatibility with clinical and radiologic picture serve as significant criteria to come up to a diagnosis. Sometimes; skin lesions are the first manifestation of systemic sarcoidosis. This is not a contagious or allergic disease. There is a risk of development of systemic manifestations at a later date; for which a close follow up is a must. We are presenting a case of cutaneous sarcoidosis, which later on progress to sarcoidosis with systemic manifestations.

Keywords: Sarcoidosis, Cutaneous, Granulomas

INTRODUCTION

Almost a century ago, the relationship of sarcoid infiltration of the skin and granulomatous changes in other organs was recognized. Schaumann in 1914, proposed that lupus pernio could be a manifestation of a generalized disease.[1] Sarcoidosis is best defined in histological terms as “a disease characterized by the presence of non caseating epitheloid cell granulomas, in several affected organs or tissues, proceeding either to a resolution or to a conversion into hyaline connective tissue”.[2] The age group more commonly affected is between 20 to 40 years, although any age group can be affected. It occurs in women twice as often. It is a granulomatous disease that commonly involves lungs, eyelids, lymph nodes and skin.[3,4.] Cutaneous sarcoidosis occurs in up to one third of patients with systemic sarcoidosis. It may have an extremely heterogeneous clinical presentation, so that the definitions of “great imitator” and “clinical chameleon” have long been used.[5] Involvement may be mild or severe, self limited or chronic, and limited or wide ranging in extent. Unfortunately, there is no single test that can prove the diagnosis. Hence diagnosis is mainly based on a compatible clinical or radiologic picture along with histologic evidence of non- caseating granulomas, and when other potential causes such as infection are excluded.[6, 7] There are no morphological features that enable the pathologist to make a diagnosis of sarcoidosis. Statements such as “consistent with sarcoidosis” or “suggestive of sarcoidosis” are helpful and may be misleading. Hence the primary role of the pathologist is (1) to identify and characterize the granulomas or document their absence[8] (2) to exclude as far as possible known causes of granulomas, primarily infections (3) to ensure compatible clinical and radiological findings. Though rare, the worst possible outcome in multisystem sarcoidosis is death due to cardiac or central nervous system damage.

CASE REPORT

A 55 year female came to MGM medical college and hospital OPD with a history of insidious onset of gradually progressive papular, erythematous lesions over the arms, back and legs over a period of 2 years. 3 months later, she developed cough and fever and gave history of weight loss. Blood investigations showed normal levels of liver function tests, kidney function tests and serum calcium. Serum angiotensin converting enzyme (ACE) levels were raised (62 micrograms/L). Multiple enlarged lymph nodes were also seen in the preaortic and para-aortic, subcarinal and aorto-pulmonary window. A skin punch biopsy was taken. Histopathological examination of the skin lesion revealed non- caseating granulomas consisting of lymphocytes and epitheloid cells and ill formed Langhan's giant cells. (Fig 1). The granulomas were seen upto deep dermis, along with a mild lymphocytic infiltrate around blood vessels and skin adnexa. (Fig 2). Biopsy stains for acid fast bacilli and periodic acid stain for fungal granulomas were negative. HRCT scan of the chest showed patchy areas of consolidation in the medial segment of the medial lobe and small calcific granuloma in the left lower lobe. (Fig 3). Thereafter, a transbronchial biopsy from the right lower and middle lobes showed small aggregates of epitheloid cells. After exclusion of infectious causes, a diagnosis of cutaneous sarcoidosis was made.

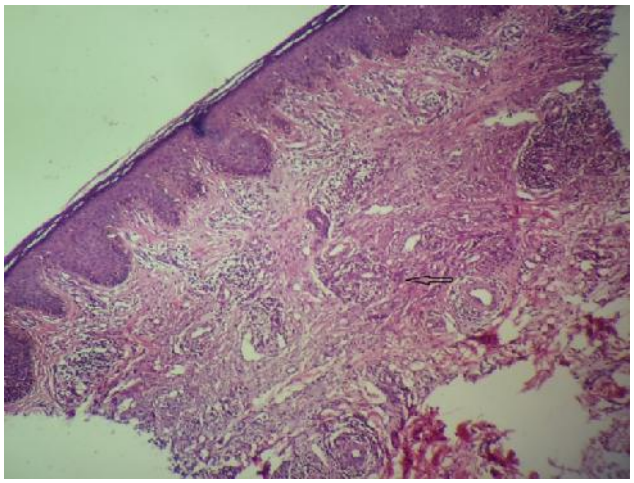

Fig 1: Depicting multiple non-caseating granulomas up to deep dermis (10X)

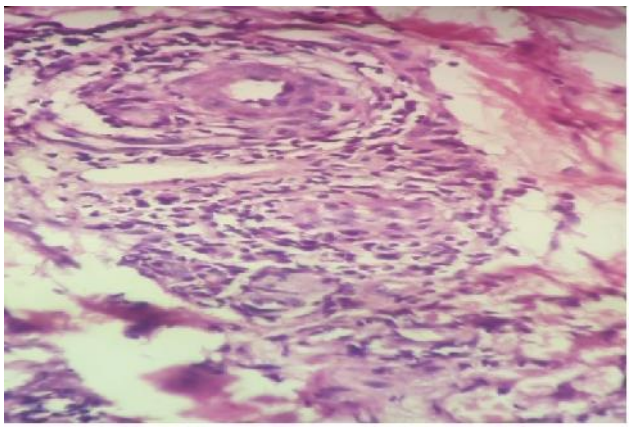

Fig 2: Granuloma with epitheloid cells, lymphocytes and ill-formed langhan's giant cell (40X)

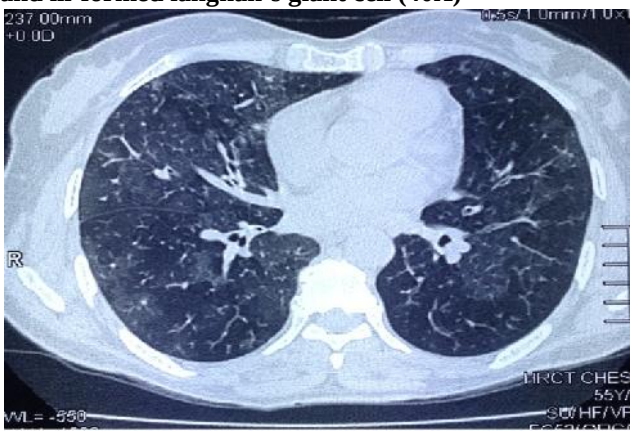

Fig 3: HRCT scan of chest showing patchy areas of consolidation in medial segment of medial lobe

DISCUSSION

Granuloma is a small, well-circumscribed lesion, 2-3 mm in diameter consisting of collection of modified macrophages (epitheloid cells) and a rim of lymphocytes. Granulomatous skin lesions present as a diagnostic challenge to dermatopathologists due to a myriad of presentations and almost identical histological pictures. A large group of skin diseases enters the differential diagnosis with cutaneous sarcoidosis. The whole word means a condition that resembles crude flesh. Several lines of evidence suggest that this disease is due to disordered immune regulation in genetically predisposed individuals. Since the clinical consequences and the prognosis of these groups of diseases is different, it is important to correctly plan the diagnostic work up. Cutaneous involvement occurs in 20% to 35% of the patients with systemic sarcoidosis. Cutaneous sarcoidosis is divided into specific and non-specific types. The most common non-specific manifestation is erythema nodosum, the biopsy of which shows panniculitis with septal inflammation. Non caseating granulomas are rarely present in erythema nodosum. The specific

skin lesions are papules, plaques, lupus pernio, subcutaneous nodules and psoriasiform lesions. Cutaneous involvement in systemic sarcoidosis may occur at any stage of the disease, however most often it presents at the onset and may even be the presenting complaint.[9] This is very true in the present case. Many atypical lesions have also been described in cutaneous sarcoidosis like itchyosiform lesions, vitiligo and scar granulomas.[10] Lung biopsy is now an established procedure in the diagnosis of radiological demonstrable pulmonary infiltration. The criteria for diagnosis of skin sarcoidosis are: 1. Clinically and radiologically compatible picture 2. Histologic evidence of non- caseating granulomas 3. Exclusion of the other granulomatous diseases like mycobacterial, fungal and parasitic infections

At the same time one should not forget non- specific local "sarcoid reaction" that also shows non-caseating granulomas, but no signs of systemic disease. Four main groups of skin conditions that mimic sarcoidosis are:- 1. Infectious diseases (Sarcoidosis is not a contagious disease) 2. Allergic and immunological manifestations of various etiologies (Sarcoidosis is not an allergic disease) 3. Granulomatous diseases of various etiologies 4. Lymphomas and pseudolymphomas

The granulomas in lupus vulgaris are caseous, those in leprosy are around dermal nerve twigs. In contrast, those in sarcoidosis are mainly in the dermis and surrounded by sparse lymphocytic infiltrate (naked tubercle). Serum angiotensin converting enzyme (ACE) levels has been used as an important laboratory test in sarcoidosis. ACE levels are derived from the epitheloid cells of the granulomas and reflect the granuloma load in the patient. It is elevated in 60% of patients, as in the present case; and is useful in monitoring the clinical course of the disease. Sarcoidosis follows an unpredictable course. 65 to 70 % of affected patients recover with minimal or no residual damage, 20 % have permanent loss of some lung function or some visual impairment, remaining 10 to 15 % die of cardiac and nervous system damage or succumb to progressive pulmonary fibrosis.

CONCLUSION

In conclusion, cutaneous sarcoidosis is present in approximately 25% of patients. Sometimes; skin lesions are the first manifestation and their recognition is important as they are an accessible source of tissue for histopathological examination. There is a risk of development of systemic manifestations at a later date; hence such patients should have a close follow up regularly. There is no permanent cure for sarcoidosis. The treatment is usually designed to help relieve the symptoms with drugs like analgesics, anti-inflammatory, steroids and chemotherapy drugs according to severity of disease.

Conflict of interest: None

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