CASE REPORT

CUTANEOUS SARCOIDOSIS

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ABSTRACT

Sarcoidosis is a multisystem granulomatous disease, still inadequately defined and it affects several systems, all of which show monotonously uniform, regular, noncaseating granulomas in all of the affected tissues. We report a case of 50 years old lady, housewife by profession presented with two erythematous annular plaque lesions on right flank and below left axilla with central atrophy since 20 years duration. Histopathological examination showed classical features of cutaneous sarcoidosis. She had no systemic involvement of disease. Cutaneous sarcoidosis has many morphological presentations and often mimics other dermatologic disease.

Key words: Cutaneous sarcoidosis, Plaques, Granulomas, Skin Biopsy

INTRODUCTION

Sarcoidosis is an idiopathic multisystem syndrome-complex. It is a granulomatous disease that commonly involves the lungs, eyes, lymph nodes and skin.[1, 2] Granulomatous skin lesions often present as diagnostic challenge to pathologists due to various modes of presentation and identical histologic picture. In Greek, Sarcoidosis means flesh-like condition. "Sarco" meaning "flesh", "eidos" meaning "like" and "osis" meaning "condition".[1] When the lesions were examined diagnosis was struck by close resemblance to sarcoma – thus the name "benign sarcoid".[3] Besnier in 1889 reported what probably was the first patient with sarcoidosis and proposed the term "lupus pernio". Hutchinson described more cases and called it "Mortimer's malady" after his famous patient. The first unequivocal case of sarcoidosis in the English Literature was reported by Boeck in 1899.[1, 3] First case in India was reported by Rajam et al. in 1957.[1, 4] Sarcoidosis has been comprehensively defined by Scadding and Mitchell as an idiopathic multisystem disease characterised by formation of non caseating epithelioid cell tubercles in affected tissues or organs. The disease process is generalised, manifestations protean and course unpredictable. [5] Between 20-35% of patients with systemic sarcoidosis have cutaneous manifestations, but cutaneous sarcoidosis can also occur without systemic disease in about 25% of cases.[5-9] Because lesions assume a vast array of morphologies, cutaneous sarcoidosis is known as one of the "great imitators" in dermatology.[1, 3, 6, 8] The lesions are both specific and nonspecific. The specific skin lesions include lupus pernio, plaques, micropapular rash, subcutaneous nodules and scars. The important nonspecific lesion is erythema nodosum. [2, 7] Diagnoses of sarcoidosis is mainly based on exclusion of other granulomatous lesions as there is no single confirmatory test which can prove the diagnosis. Patients are diagnosed as sarcoidosis when a compatible clinical or radiologic picture is present along with histologic evidence of noncaseating granulomas, and when other potential causes, such as infections, are excluded. [6]

CASE HISTORY

A 50 year old female presented with insidious onset of gradually progressive, red plaque over right flank. The plaque showed annularly active lesion and central atrophy. Same kind of lesion appeared on left side below the axilla. The size of lesion on right side is 15X15cm and on left side 12X12 cm. There was no history of fever, cough, joint pain, eye complaints, weight loss or any other systemic complaints. There was no history of trauma. General physical and systemic examinations were normal. Past and family history was not contributory. There was no significant drug history. All routine hematological investigations were normal. Chest X ray showed mild increase in bronco vascular markings. There was no evidence of hilar lymphedenopathy. Sputam AFB was negative. Serum Angiotensin converting enzyme (ACE) titer was 48.5 U/L. Serum calcium, liver function tests(LFT), renal function tests (RFT)and ultra sonogram (USG) abdomen was normal.

Dermatological examination reveled two well defined non tender, smooth flat topped hypopigmented plaques with erythematous nodules annularly and

central atrophy. There was no loss of sensation. [Figure 1]

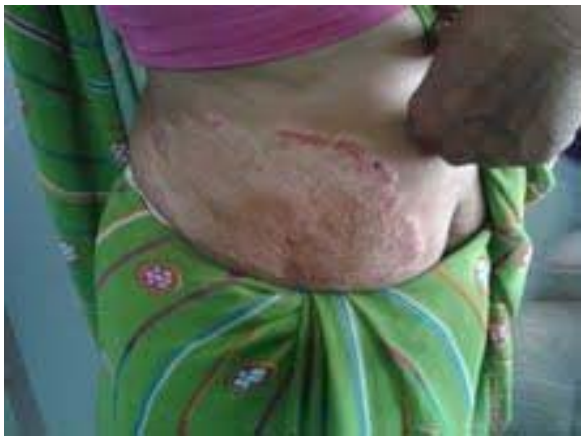

Figure 1: Hypopigmented plaque with annular erythmatous lesions

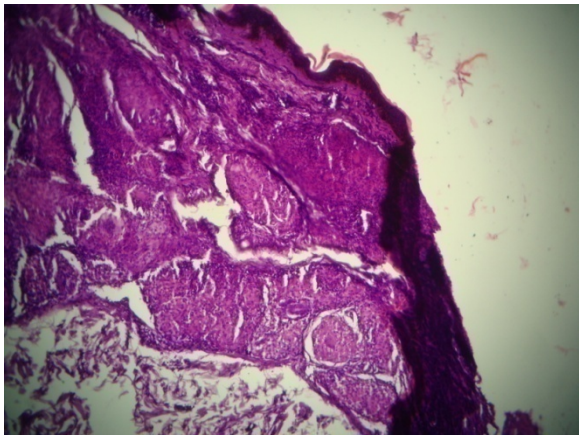

Figure 2: Photomicrograph showing granulomatous infiltrate in the dermis. (H & E stain, 20X)

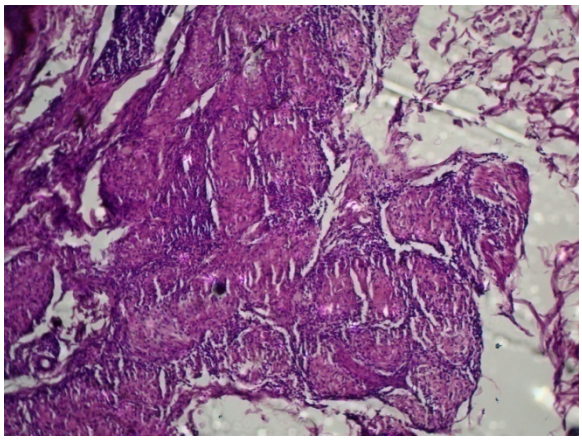

Figure 3: Photomicrograph showing the close up of the granuloma (H & E stain, 40X)

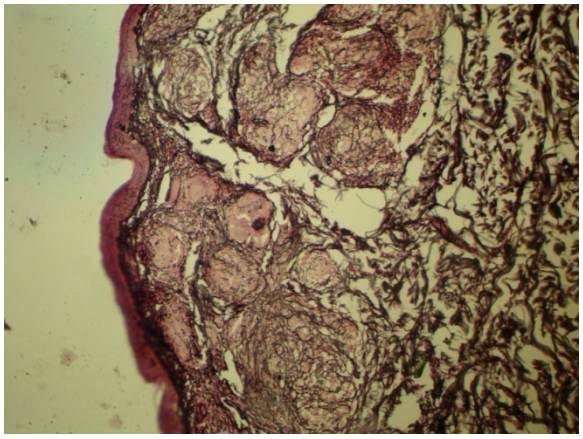

Figure 4: Reticulin stain showin intect reticulin fibers in and around the granulomas (Reticulin stain, 20X)

Histopathological examination of the skin lesion showed well defined noncaseating epitheliod granulomas and mild infiltrate of lymphocytes arranged in a band like pattern in the dermis. [Figure2] The granulomas are composed of epitheloid cells, langhans type of giant cells and few lymphocytes. No caseous necrosis seen. [Figure 3] Biopsy stains for acid fast bacillus And Periodic acid stain for fungal granuloma were negative.

Reticulin stain revealed reticulin fibers in around the granulomas. [Figure 4] Skin biopsy was suggestive of non caseating granulomatous lesion and diagnosis of Cutaneous Sarcoidosis was made. Patient underwent treatment with local and systemic Steroid therapy.

DISCUSSION

Granulomatous skin lesions often present as a diagnostic challenge to dermatopathologists due to various modes of presentations and identical histological pictures. Sarcoidosis is the result of an immune dysfunction due to a persistent antigen of low virulence that is poorly cleared by the immune system.[5] Sarcoidosis is a multisystem disorder which involves lungs, eyes, skin, reticuloendothelial system, liver, salivary glands, bones and almost any organ in the body.[1] Granulomas are relatively discrete collections of histiocytes or epitheliod histiocytes with variable number of admixed multinucleated giant cells and inflammatory cells.[10] Sarcoidosis occur worldwide with the highest prevalence in Scandinavia and the prevalence was 20/1, 00, 000 in 1975 but recent report says it to be 64/1, 00, 000.[1] only 300cases of sarcoidosis were reported in our country till 1996 but a recent report says there is an increased prevalence.[1] The frequency of cutaneous involvement varies considerably, from 11 percent to 90 percent.[7] In one study [11] approximately 30% of of patients who initially had only cutaneous lesions develops systemic involvement months to years later. Sarcoid is a great

imitator of other dermatologic diseases because the cutaneous manifestations are quite variable and occur in both localized as well as generalized forms. [8] The relationship between cutaneous and systemic sarcoidosis is being studied. Cuta-neous involvement in systemic sarcoidosis may occur at any stage of the disease. However, it is most often present at the onset and may even be the presenting complaint. Every patient with cutaneous sarcoidosis requires an initial work-up for systemic involvement, followed by periodic screening. [6] Cutaneous lesions of sarcoidosis are classified into non specific and specific types. Non specific manifestations present as erythema nodosum, erythema multiformae, calcinosis cutis or nummular eczema. Specific types are classified as maculopapular, papular (lichenoid), nodular (annular, angiolupoid, subcutaneous), plaque (lupus pernio) and erythematous types depending on the type and extent of involvement of the skin and subcutaneous tissues. [5] Many atypical skin lesions have been described in sarcoidosis such as extensive ulcerative lesions, psoriasiform plaques, hyperpigmentation in black patients, verrucous and papillomatous lesions, ichthyosiform lesions, pustular folliculitis, papules in light exposed areas, lichenoid eruptions, erythrodermic eruptions, cicatrical alopecia, lupus erythematosus- like lesions, mutilating lesions, erythema and plaques involving pams and soles, pruritus and diffuse skin plaques.[3, 5]

The diagnosis of skin sarcoidosis depends upon the following criteria: (1) compatible clinical or

radiologic picture or both; (2) histologic evidence of noncaseating granulomata; (3) negative cultures (acid-fast bacilli, fungi) of sputum and biopsied tissue. The diagnosis is open to misinterpretation if all the three criteria are not satisfied. The presence of noncaseating granulomas alone without corroborative laboratory evidence may be due

to a nonspecific local sarcoid reaction for which there are many causes. Conversely, without the histologic confirmation, the differential diagnosis of clinical syndrome is extensive and includes various bacterial, viral, and fungal disorders.[7] Sarcoidosis needs to be distinguished histopathologically from lupus vulgaris and leprosy as they all have epithelioid cell granulomas. While the granulomas in lupus vulgaris are caseous and present in the upper dermis, those in leprosy are mainly around dermal nerve twigs and admixed with abundant lymphocytic infiltration. In contrast, sarcoidal granulomas are discrete, distributed uniformly in the dermis and surrounded by sparse lymphocyte cuffing ('naked tubercles'), with fine reticulin fibers in and around the tubercles.[5]

Serum level of Angiotensin converting enzyme (**ACE**) has been used as an important laboratory test in sarcoidosis since 1975. However, Bunting et al reported that increased serum ACE level is not specific for sarcoidosis because the sensitivity and specificity were 77% and 93% respectively. In our case, , the serum ACE level was normal, indicating that the level is not associated with the disease activity. The volume of sarcoidal granulomas in the lichenoid type of sarcoidosis may be too small to produce a high level of ACE to be detected as an elevated serum level. [2] ACE level in serum derived from epithelioid cells of the granulomas and reflects the granuloma load in the patient. ACE levels are neither diagnostic nor predictor of systemic involvement. It is elevated in 60% of patients and is useful in monitoring the clinical course of the disease. [5] Steroid treated cases have near normal value of A C E. It was found useful to determine the activity of disease.

To conclude, recognition of cutaneous sarcosid lesions is very important because they provide an important visible clue to the diagnosis and are an accessible source of tissue for histopathological examination. Cutaneous sarcoidosis has been reported occasionally from India and in most of these reports; lesions were associated with some from of systemic involvement. Therefore patient presented with disease confined to skin alone, should be followed-up regularly for the probable risk of developing systemic manifestations at a later date.

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