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Case Report

Cutaneous sarcoidosis: a marker of underlying systemic disease

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ABSTRACT

Sarcoidosis/Mortimer's malady is a multisystem disease defined by presence of epithelioid cell granulomas without caseation in various organs. It involves mainly the lungs, eyes, skin, lymph node etc. We reported a 40 year old female who presented with multiple plaques over different regions of the body with past history of ulcerated lesion of scalp which healed with cicatricial alopecia. On thorough evaluation she was found to have involvement of multiple internal organs. Based on the clinical features and histopathological findings, a diagnosis of cutaneous sarcoidosis (plaque type) was made. Patient treated with systemic and topical corticosteroids resulting in marked improvement in her skin lesions were seen and pulmonary complaints also resolved.

Keywords: Plaque sarcoidosis, Lung fibrosis, Mortimer's malady

INTRODUCTION

Sarcoidosis/Mortimer's malady is a multisystem disease defined by presence of epithelioid cell granulomas without caseation in various organs.[1] It involves mainly the lungs, eyes, skin, lymph node etc.[2] Skin involvement is seen in 20%-35% of patients with sarcoidosis and cutaneous lesions are frequently the presentation of the disease in 1/3rd of the patients.[3]

We reported a case of cutaneous sarcoidosis who presented with multiple plaques over different regions of the body with past history of ulcerated lesion of scalp which healed with cicatricial alopecia. On thorough evaluation she was found to have involvement of multiple internal organs.

CASE REPORT

A 45 year old female patient presented with a chief complaint of raised lesions over face, trunk, and limbs since 3 months. Initially patient developed a single solid raised lesion over mid lower back which gradually increased in size up to 3 cm. She later developed multiple similar lesions of various sizes over adjacent areas of back, upper limbs, lower limbs and face. Lesions are associated with pruritus. She also complained of mild shortness of breath. Past history revealed that patient had painful pus filled lesions over the scalp 2 years back which was diagnosed as Lupus vulgaris of scalp for which she used anti tubercular treatment for a period of 12 months. Few areas of cicatricial alopecia were present over scalp. Patient is a known diabetic and on regular medication.

On dermatological examination multiple skin coloured infiltrated plaques over forehead and bilateral cheeks were seen. Some of the plaques over the face showed central clearing and annular configuration (Figure 1). Multiple closely arranged erythematous well defined scaly papules and plaques were seen over back, proximal extremities. Post-inflammatory hyperpigmentation was seen over some healed areas (Figure 2).

Based on history and clinical examination a differential diagnosis of borderline lepromatous leprosy, sarcoidosis, discoid lupus erythematous, psoriasis and polymorphic light eruption were considered. Complete blood picture, serum biochemistry were normal except for raised random

blood sugars. Serum calcium was within normal limits. Slit skin smear taken from ear lobes and skin lesions were negative for acid fast bacilli.

Electrocardiogram revealed no abnormality. A 4 mm skin sample for histopathological examination was sent. Histopathology showed stratum corneum with parakeratosis, diminished stratum granulosum. Stratum spinosum was reduced in thickness and showed complete effacement of rete ridges. In addition, few vacuolated (necrotic) keratinocytes were noted. The superficial dermis showed granulomatous infiltrate composed of epithelioid histiocytes (Figure 3), foreign body as well as langhan's type of giant cells and sparse lymphocytes. The granulomas were small, naked and confluent. The deep dermis also showed few granulomas. In view of small, naked and confluent granulomas, the possibility 'cutaneous sarcoidosis' was considered. Based on the clinical features and histopathological findings, a diagnosis of cutaneous sarcoidosis (plaque type) was made.

Patient was further thoroughly evaluated and investigated to detect any systemic involvement secondary to sarcoidosis. Imaging studies were performed. Chest X-ray revealed no evidence of hilar lymphadenopathy. High resolution computed tomography showed few tiny perilymphatic micronodules in anterior segment of right upper lobe and apico-posterior segment of left upper lobe with adjacent ground glass opacity. Small focal areas of fibrosis with adjacent ground glass attenuation and traction bronchiectasis were seen in posterior basal segments of lower lobes. Few small dense fibrotic areas seen in apicoposterior segments of both upper lobes (Figure 4).

In mediastinum, few small lymph nodes seen in prevascular, para aortic and pre tracheal regions (short axis diameter less than 5 mm). The above findings were consistent with pulmonary sarcoidosis. Thyroid function test showed hypothyroidism with raised TSH of 10.5 mU/l. Serum angiotensin converting enzyme is markedly elevated (605.4 u/l). Mantoux showed no induration. On ophthalmic examination no evidence of uveitis was seen but schirmer's test is positive with grade-1 abnormality (wetting of 10- 15 mm of a schirmer's strip indicating mild dry eyes).

Patient was treated with 30 mg oral prednisolone once daily for 4 weeks with gradual tapering over 8 weeks. Topical betamethasone dipropionate cream twice daily for 4 weeks. Oral hydroxy choloroquine 200 mg/day along with multi vitamin supplements were also given.

Marked improvement in her skin lesions was seen within 4 weeks and her pulmonary complaints also improved markedly (Figure 5).

Patient was on regular follow up.

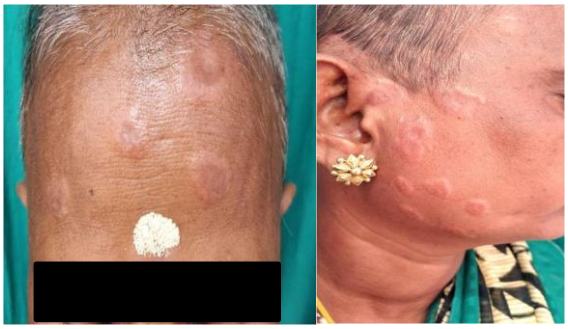

Figure 1: Multiple skin coloured infiltrated plaques over forehead and bilateral cheeks. Some of the plaques showed central clearing and annular configuration.

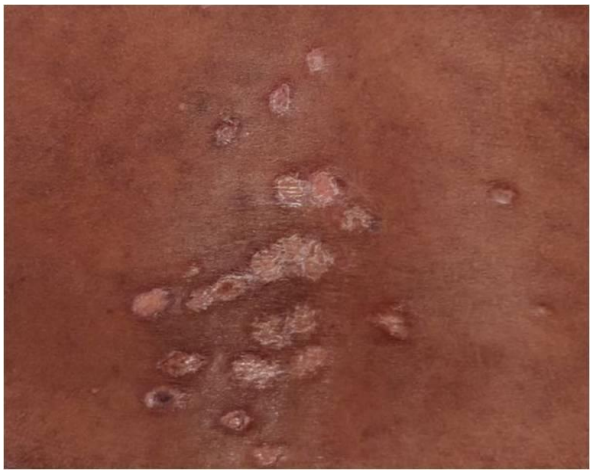

Figure 2: Multiple closely arranged erythematous well marginated scaly papules, plaques were seen over upper and lower back and proximal extremities. Post-inflammatory hyperpigmentation was seen over some healed areas.

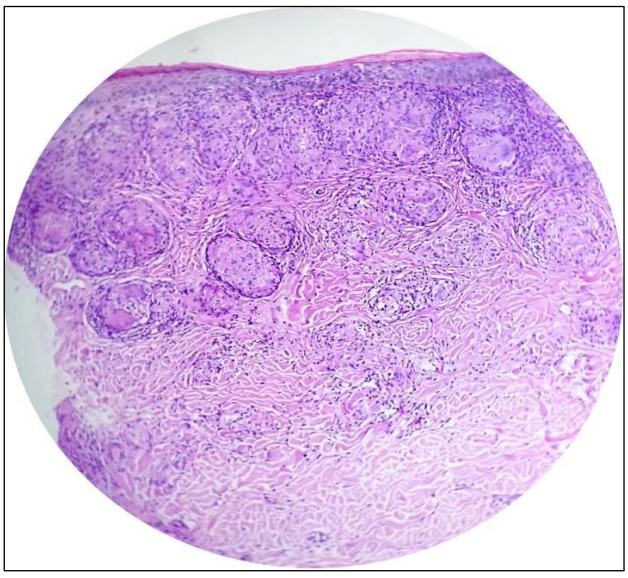

Figure 3: H and E section low power view- multiple discrete epithelioid granulomas.

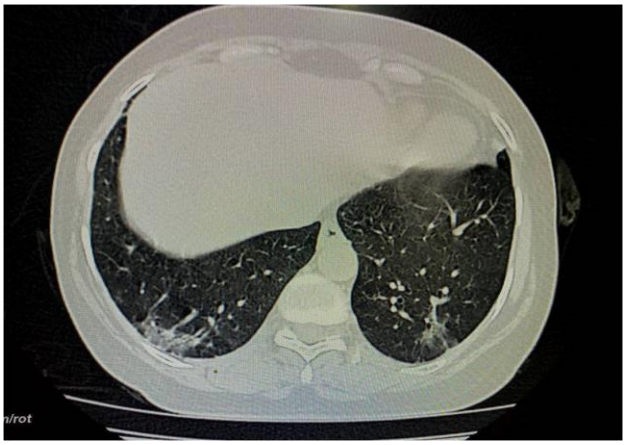

Figure 4: Small focal areas of fibrosis in posterior basal segments of both lower lobes.

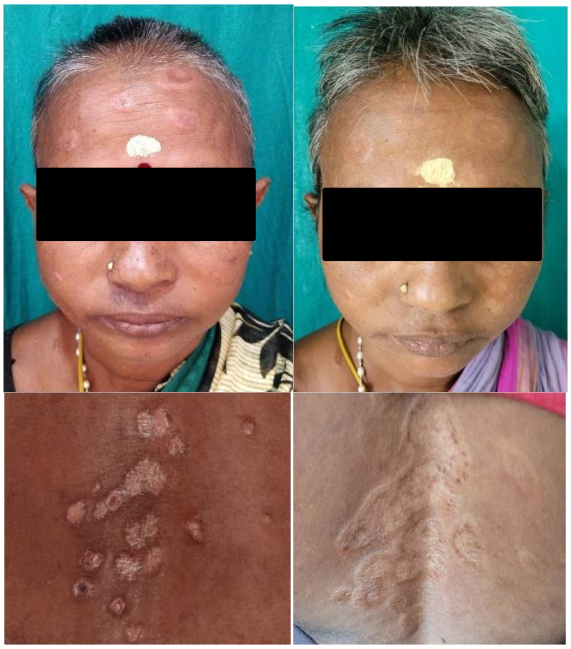

Figure 5: Lesions before and after treatment.

DISCUSSION

Sarcoidosis commonly affects the lungs, mediastinal and peripheral lymph nodes, liver, eyes, bones, and skin. Hutchinson recorded the first case in 1865 and called it Mortimer's Malady after his famous patient Mrs. Mortimer.[4] Cutaneous lesions of sarcoidosis may be specific or non-specific. As the lesions assume a vast array of morphologies, cutaneous sarcoidosis is considered as one of the 'great imitators' in dermatology.[5] The specific forms of cutaneous sarcoidosis include maculopapular, nodular, plaque, subcutaneous and scar sarcoidosis, lupus pernio, and certain less common forms include psoriasiform, lichenoid, hypopigmented, erythrodermic, verrucous, and morphea-like.[6,7] Non-specific forms include erythema nodosum and rarely erythema multiforme, calcinosis cutis, and prurigo. Histopathologically, it is characterized by naked granulomas which are aggregates of epitheloid cells with a sparse lymphocytic component, a few langhan's giant cells and inclusion bodies namely schaumann/asteroid/residual bodies.[8] Multi-system involvement is characteristic of the disease. Pulmonary involvement occurs in 90% of the cases of sarcoidosis.[9] Skin lesions appearing before systemic disease is known to occur in 20% of cases. However, in this case, the cutaneous lesions guided the way to identify the lung, eye and thyroid involvement. In the present case papulo nodular and plaque forms were seen. Based on her clinical features and extensive involvement patient was subjected to investigations which revealed underlying systemic involvement. Intra thoracic involvement occurs in 90% of case.[5]

At presentation, mid and upper zone pulmonary opacities (i.e., nodules and reticulation) are seen in 20-50% of patients. The nodules vary in size and can coalesce and cause alveolar collapse, thus producing consolidations. The classic pattern on high resolution computed tomography is widespread, well-defined nodules with smooth or irregular margins measuring 2-5 mm in diameter, present in 80-100% of cases, even in patients with atypical radiological presentations. Pulmonary fibrosis is seen in 20-25% of pulmonary sarcoidosis, has upper lobe predilection, with distortion of lobar architecture and volume loss with or without honeycombing. In the present case few tiny peri-lymphatic micronodules were seen in anterior segment of right upper lobe and apico-posterior segment of left upper lobe with adjacent ground glass opacity. Small focal areas of fibrosis with adjacent ground glass attenuation and traction bronchiectasis were seen in posterior basal segments of lower lobes.

A combination of more characteristic findings on imaging can be diagnostic of sarcoidosis, for example, peri-lymphatic micronodules with upper lobe distribution with bilateral symmetrical hilar and subcarinal lymphadenopathy. The present case had few tiny peri-lymphatic micronodules are seen in anterior segment of right upper lobe and apico-posterior segment of left upper lobe. Ocular manifestations may be the presenting sign of sarcoidosis in 11-30% of patients. Any segment of the eye and its adnexa may be affected, but uveitis is the most frequently diagnosed condition followed by chorioretinitis, periphlebits, papilloedema, lacrimal gland involvement, kerato conjunctivitis sicca.[13] In the present case ophthalmological examination showed dryness of eyes and no evidence of uveitis was noticed.

Thyroid infiltration by sarcoidosis was first described in 1938 and was reported to be approximately 4% in autopsy series. Thyroid dysfunction has been reported in 16%-30% in various series, which is predominantly hypothyroidism.[14] In the present case serum TSH was elevated. Patient was treated with L-thyroxine of 50 µg/day. Cutaneous sarcoidosis is treated with topical or intralesional (i.e.; injected) corticosteroids as the first-line therapy for localised and mild disease. Systemic

treatments are used for cutaneous sarcoidosis primarily when it does not respond to first-line therapies. These include oral corticosteroids, chloroquine or hydroxychloroquine, methotrexate, the tetracycline class of drugs, thalidomide, and tumour necrosis alpha inhibitors, such as infliximab. In view of significant pulmonary involvement the present case was treated with oral corticosteroids and hydroxychloroquine which led to significant clinical improvement with no side effects to any of the drugs.

CONCLUSION

Considering the fact that cutaneous sarcoidosis is often a precursor of the systemic form, thorough evaluation should be conducted to determine systemic involvement. Prompt detection and treatment of systemic involvement helps in preventing long term morbidity and complications.

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