ORIGINAL RESEARCH PAPER
Volume - 11 | Issue - 02 | February - 2022 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

SKIN LESION – A WINDOW TO A SYSTEMIC DISEASE

Dermatology

Nithila M* Postgraduate, Department of Dermatology, Sri Ramachandra Institute of Higher Education and Research. *Corresponding Author

Adikrishnan S Professor, Department of Dermatology, Sri Ramachandra Institute of Higher Education and Research.

Murugan S Professor, Department of Dermatology, Sri Ramachandra Institute of Higher Education and Research.

ABSTRACT

Sarcoidosis is considered as one of the "great masqueraders" in the field of dermatology. Polymorphic cutaneous lesions are seen in around 20- 35 % of patients with sarcoidosis. We report a 49 year old male who presented with cough and hypopigmented atrophic plaques over the back. Serum ACE levels and CRP were elevated. HRCT chest revealed parenchymal and hilar, mediastinal lymph node involvement. Skin biopsy was done and showed multiple naked non- caseating granulomas in upper dermis. Pulmonary function test was suggestive of peripheral obstructive airway disease. Bronchoalveolar lavage identified CD4+ lymphocytosis. The patient was started on topical tacrolimus and topical corticosteroids with which he showed improvement. The patient was referred to pulmonology for further management. The case is presented for its rarity, with skin manifestations being one of the presenting signs of the disease.

KEYWORDS

Sarcoidosis, atrophic, granulomas

INTRODUCTION

Sarcoidosis is a systemic disease of unknown etiology. It is a granulomatous disease which is characterised by infiltration of organs with non-caseating granulomas. The most commonly affected organs are lung, lymph nodes and eyes. The term 'Mortimer's Malady' was coined by Jonathan Hutchinson in the year 1898.Cutaneous involvement is usually polymorphic and it is seen in around 20-35% of patients having systemic involvement[1]. Among these cases, exclusive cutaneous manifestation is limited to 10% of patients[2].

CASE REPORT

A 49 year old male presented with cough and a single episode of expectoration of blood stained sputum along with asymptomatic multiple whitish skin lesions over the back for the past 2 months. On examination, the patient had multiple ill-defined hypopigmented atrophic plaques of varying sizes from 0.25 cm x 0.25 cm to 1 cm x 1 cm over the centre of his back as shown in Fig 1. The clinical differentials for the skin lesions considered were Lichen sclerosus et atrophicus, early morphoea and sarcoidosis. A skin biopsy of the lesion revealed non-caseating naked granulomas in the dermis consisting of epithelioid cells and Langhans giant cells as shown in Fig 2,3. Blood investigations revealed low hemoglobin (11.5 g/dl), microcytic hypochromic anemia in peripheral smear, normal serum calcium (9.5 mg/dl), elevated ACE(83.53 U/L) and CRP(13.5 mg/L). Mantoux revealed 5 mm induration. HRCT chest revealed multiple nodular opacities in the perilymphatic distribution in bilateral upper lobe, right middle lobe and superior segment of bilateral lower lobes as shown in Fig 4. Patchy pleural based consolidation with few subpleural nodules in the posterior basal segment of the left lower lobe and multiple discrete non-calcified nodes in bilateral hilum and mediastinum were also noted. Pulmonary function test revealed features suggestive of peripheral obstructive airway disease. Bronchoscopy was done and the specimen was negative for AFB and culture showed no growth. Bronchoalveolar lavage showed $CD4^{+}$ lymphocytosis. The patient was started on topical tacrolimus and topical corticosteroids following which there were resolution of lesions. The patient was referred to Pulmonology department for further management.

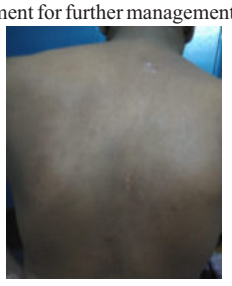

Fig 1: Hypopigmented atrophic plaques over back

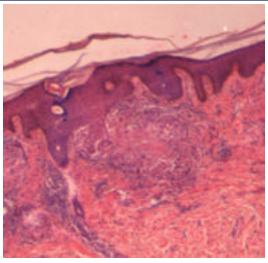

Fig 2: Histopathological examination shows naked granulomas in upper dermis(100X)

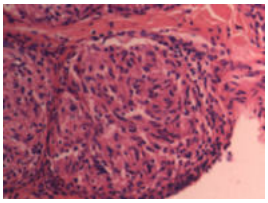

Fig 3: Histopathological examination showing the presence of epithelioid cells and Langhans giant cells(400X)

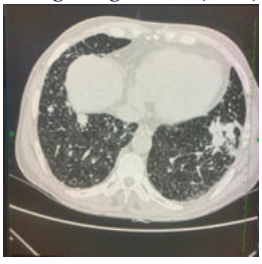

Fig 4: HRCT showing pulmonary involvement

DISCUSSION

Sarcoidosis, which is also known as Besnier-Boeck-Schauman disease, was first described by an english doctor Jonathan Hutchinson[3].It is usually seen in the third and fourth decades. Sarcoidosis is an antigen mediated disease of unknown etiology. It is thus a diagnosis of exclusion[4].The cutaneous lesions of sarcoidosis present with varying morphologies and hence Sarcoidosis is considered as one of the "great masqueraders" in the field of dermatology. It is hypothesized that exposure to one or more extrinsic antigens in an individual who is genetically susceptible leads to inflammatory pathways' reactivation which facilitates the formation of granulomas[5]. Cutaneous lesions can either be specific, showing

sarcoid granulomas histopathologically or non-specific lesions which show no granulomas on biopsy[6].The most common specific lesions are maculopapules, plaques, lupus pernio, scar-sarcoidosis and subcutaneous sarcoidosis. The most important non-specific lesion is Erythema nodosum. Other less common forms of sarcoidosis include hypopigmented, psoriasiform, lichenoid, verrucous, ulcerative, necrobiosis lipoidica-like lesions, angiolupoid, ichthyosiform, erythrodermic, morphoea-like lesions and livedo. An evaluation for systemic sarcoidosis includes a complete history and physical examination, baseline laboratory testing (calcium, renal function, hepatic function), ACE levels, chest radiography, pulmonary function testing, electrocardiography, ophthalmologic evaluation, tuberculin skin test and biopsy(including culture for mycobacteria and fungus). Elevated ACE levels are seen in approximately 50-60% of patients with Sarcoidosis[5]. Histopathological examination of the biopsy specimen reveals non-caseating naked granulomas composed of epithelioid cells and multinucleate giant cells. The granulomas are referred to as naked because the lymphocytic infiltrate at the margins of the granuloma is very sparse. Schaumann bodies, asteroid bodies, Hamazaki-Wesenberg bodies, and calcium oxalate crystals[7],[8] are also seen in the biopsy specimens. Topical, intralesional corticosteroids and topical tacrolimus are used for localized lesions. Systemic management includes systemic corticosteroids, Methotrexate, Antimalarials, Tetracycline and TNF-alpha inhibitors for recalcitrant cases.

CONCLUSION

The case is presented for its rarity and as in this case, skin manifestations has been one of the presenting signs of systemic sarcoidosis. Morphoea-like lesions, as seen in this patient is a rare presentation of cutaneous sarcoidosis. The case emphasizes the fact that cutaneous lesions in Sarcoidosis represent only the tip of the iceberg. Lung involvement is seen in around 90-95% of cases and hence patients with cutaneous sarcoidosis require a thorough evaluation[5]. A sound knowledge of cutaneous sarcoidosis is vital for early diagnosis and intervention of systemic sarcoidosis.

REFERENCES

1. Pandhi D, Sonthalia S, Singal A. Mortimer's Malady revisited: A case pf polymorphic cutaneous and systemic sarcoidosis. Indian J Dermatol Venereol Leprol 2010; 76: 448
2. James WD, Berger TG, Elston DM. Sarcoidosis.Andrews' diseases of the skin, 10th ed. Canada: Saunders elsevier; 2006. 708-14.
3. James DG, Sharma OP. "From Hutchinson to now: a historical glimpse" . Current Opinion in Pulmonary Medicine; September 2002. 8 (5): 416–23.
4. Myers JL, Tazelaar HD. Challenges in pulmonary fibrosis: Problematic granulomatous lung disease. Thorax. 2008;63:78–84.
5. Haimovic A, Sanchez M, Judson MA, Prystowsky S (2012). Sarcoidosis: A comprehensive review and update for the dermatologist: Part I. Cutaneous disease. , 66 (5), 699.
6. Mana J, Marcoval J, Graells J, Salazar A, Peyri J, Pujol R. Cutaneous involvement in sarcoidosis. Relationship to systemic disease. Arch Dermatol. 1997;133:882–8.
7. Judson M.A. The diagnosis of sarcoidosis. Clin Chest Med. 2008;29(3):415–427.
8. Govender P., Berman J.S. The diagnosis of sarcoidosis. Clin Chest Med. 2015;36(4):585–602.