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Cutaneous Sarcoidosis- An Enigma And A Great Imitator In Dermatology: A Case Series.

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ABSTRACT

Sarcoidosis is an idiopathic multisystem disease characterized by non-caseating granuloma formation in the affected organ. Isolated skin involvement without other organ involvement is quite rare. Cutaneous Sarcoidosis presents with a wide variety of signs and symptoms, mimicking various other common dermatologic abnormality which causes diagnostic confusion. We have reported a series of four cases of Cutaneous Sarcoidosis over a period of three years. Cutaneous Sarcoidosis is a great imitator of other dermatological diseases and early accurate diagnosis is essential for proper management of the patients.

Keywords: Cutaneous Sarcoidosis, noncaseating granuloma, idiopathic multisystem disease

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INTRODUCTION

Sarcoidosis is an idiopathic multisystem granulomatous disease.The first case of Sarcoidosis was described by Jonathon Hutchinson, which was a dermatological curiosity of that time [1]. With the progress of research in medical science, it gradually evolved into a multisystem disease [2]. Global incidence of Sarcoidosis is roughly 16.5 per 100000 in male and 19 per 100000 in female [3]. Incidence in female (2.4%) is greater than male (1.3%) [4]. The organ commonly involved are lung, lymph node, eye, skin. The disease commonly begins at around 40 years and more common in female [5]. Histopathologically it is characterized by presence of noncaseating granulomas in the organ involved. Sarcoidosis confined to skin is quite uncommon. Here we have reported a series of four cases of Cutaneous Sarcoidosis over a period of three years.

MATERIALS AND METHODS

The study was conducted in a tertiary care hospital over a period of 3 years. In this case series punch biopsy skin specimens were received in Pathology department, processed by routine tissue processing method& paraffin blocks were prepared. Then histopathological Slides were prepared, stained by Hematoxylin and Eosin stain and examined under microscope.

The other common causes of granulomatous disease such as TB, Leprosy were excluded by ZN stain & Wade Fite stain respectively before to confirm the cases as Cutaneous Sarcoidosis.

Necessary clinical data, such as- site and nature of the lesion, laboratory parameters such as Complete blood count (CBC), chest X-ray, serum Angiotensin Converting Enzyme (ACE) level were collected from clinical records.

Consent Form

Informed written consent form taken from each patient who were included in this study.

Ethical Approval

This study was approved by institutional ethical committee.

RESULTS

Case Description

Case 1- A 48 yrs, female presented with red maculopapules over face, predominantly in nasolabial fold and around orbit.

Case 2- A 23 yrs female presented with papular lesion over left knee with history of repetitive trauma and scar over that site.

Case 3- A 54 year male presented with plaque over occipital aspect of scalp and neck. The neck lesions are violaceous in colour.

Case 4- A 46 year female presented with itchy erythematous lesion over face and extremities. Lesions were associated with scaling.

In all cases, there was no suggestive history of systemic disease. General and systemic examination were unremarkable.

Findings

Complete blood count (CBC), chest X-ray, LFT, USG abdomen were unremarkable. Serum Angiotensin Converting Enzyme(ACE) were between 60U/L to 120 U/L.

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Histopathological examination of skin tissue showed presence of multiple granulomas, predominantly composed of epithelioid cells, with sparse lymphocytes. ZN smear were negative for Acid fast bacilli in all the cases. 3 out of 4 cases were female and 3 cases were between 40 to 60 years age group. Most common site involved was face and most common presentation was erythematous papular lesion.

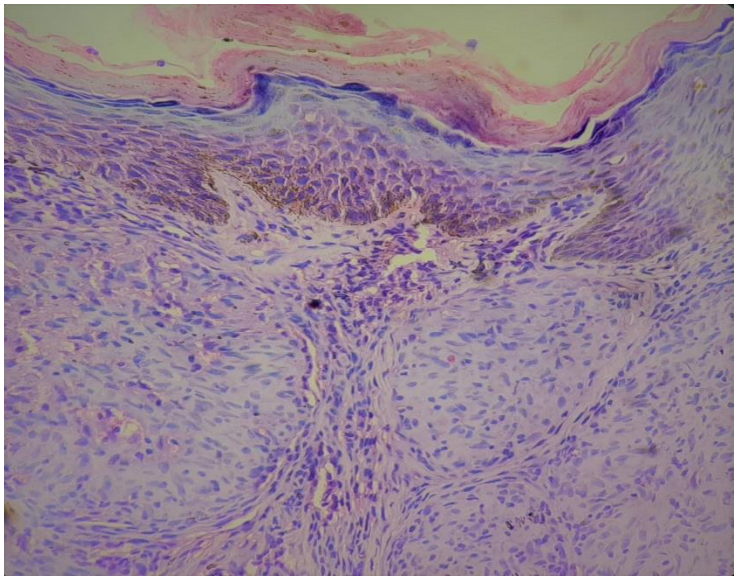

Figure 1: Showing multiple granulomas in the dermis containing predominantly epithelioid cells with sparse lymphocytes (Hematoxylin & Eosin stain, High powerobjective)

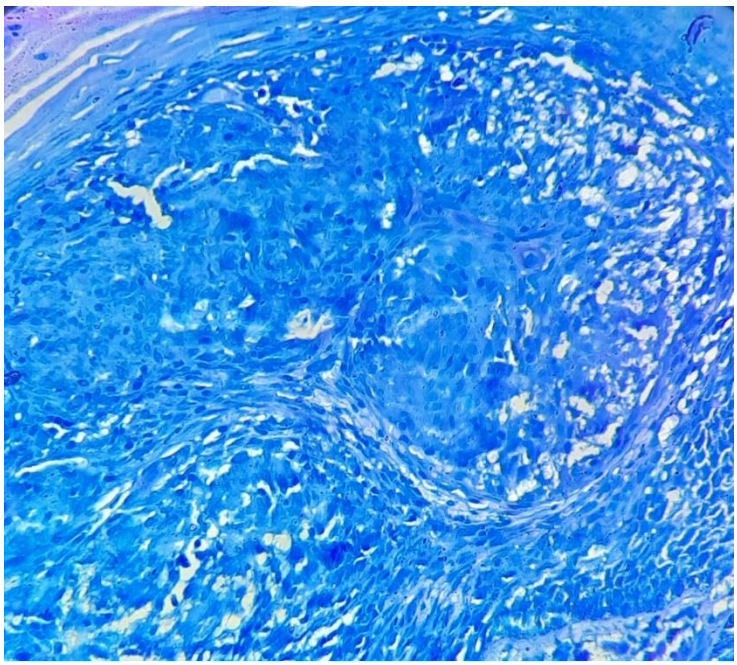

Figure 2: Showing absence of AFB (Acid fast bacilli) in the corresponding histopathological sections (Ziehl Neelsen Stain , Oil immersion Objective)

DISCUSSION

Sarcoidosis is an idiopathic multisystem granulomatous disease commonly involving lung, lymph node, eye, skin [5]. It affects people all over the world .The first case of Sarcoidosis was described by Jonathon Hutchinson. The first case in India was reported by Rajan et al in 1957 [5]. Till 1986, 300 cases

were recorded in our country. But with better understanding of diseases and newer investigation modalities, more cases were diagnosed in recent times [6]. Involvement of skin in cases of systemic sarcoidosis is between 20-35% [6]. Isolated cutaneous involvement in Sarcoidosis is uncommon and seen in around 10% of cases [7].

The disease is usually seen in middle aged population(begins at around 40 years of age) and two thirds of cases are female [8]. In our study 3 out of 4 cases were female and 3 cases were between 40 to 60 years age group.

Cutaneous Sarcoidosis is rare in India [9]. In our study, we have reported four cases of Cutaneous Sarcoidosis.

Cutaneous Sarcoidosis is known as great imitator in dermatopathology because of its wide variety of clinical presentation such as- papule, maculopapule, plaque, scar sarcoidosis, subcutaneous nodule, lupus pernio, erythema nodosum, ulcer, alopecia [10]. Although papule is the commonest presentation, colour of papule may vary such as- red, reddish brown, violaceous, translucent to hyperpigmented. These feature contribute to the entity of Cutaneous Sarcoidosis to be known as great masquerader [5].

Case1- It was a case of typical Cutaneous Sarcoidosis with common clinical presentation of involving middle aged female presenting with maculopapular lesion over face.

Case 2- It was a case seen in 23 year female, which is a relatively uncommon age group of Cutaneous Sarcoidosis. The clinical presentation was involvement of skin over knee with scar with history of repeated trauma. This was a case of Scar Sarcoidosis. Cutaneous Sarcoidosis lesion can appear on pre-existing scar [10]. A scar may develop due to injury, surgery, burn or tattoo. According to study by Naville et al , 29% of cases of Cutaneous Sarcoidosis had Scar Sarcoidosis [11].

Case 3- The lesions were violaceous in colour, which was a rare finding. It was a great mimic of common clinical condition Lichen Planus and case was diagnosed as Lichen Planus clinically. However, on histological examination, it was diagnosed as a case of Sarcoidosis.

Case 4- Patientpresented with itchy erythematous lesion associated with scaling. In approximately 10-15% of cases with Cutaneous Sarcoidosis may itch [7]. Itchy erythematous lesion with scaling represent psoriasiform lesion. Psoriasiform lesion rarely reported as Cutaneous Sarcoidosis, as stated by Burgoyne et al [12]. We reported four cases of Cutaneous Sarcoidosis, with wide variety of clinical presentation.

CONCLUSION

Cutaneous Sarcoidosis, due to its wide range of clinical presentation, is a great imitator of other dermatological condition. Greater awareness and understanding of wide range of clinical manifestation could help to prevent misdiagnosis of Sarcoidosis as well as to prevent diagnostic delay and it would help in initiating appropriate therapy of the patients with better management.

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Contribution From The Authors

- Conceptualization: Dr Rajashree Pradhan
- Project administration: Dr Sajeeb Mondal
- Supervision: Dr Ashmita Chakraborty
- Writing - original draft: Dr Rajashree Pradhan
  Writing - review & editing: Dr Sankha Chatterjee

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