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A Middle Aged, South Asian Woman with Oid Oid Disease: A Case Report

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Abstract: Described in 1937, Sulzberger Garbe dermatosis is a rare dermatological disease entity with not more than 100 cases in reported literature. The disease has mainly been reported in middle aged Jewish males but can be seen in all age groups. The patients mainly present with discoid exudative and lichenoid plaques that are often associated with severe pruritus and are usually found on the trunk and extremities. Eosinophilia is usually found.

Keywords: Sulzberger Garbe dermatosis, discoid, lichenoid plaques, eosinophilia

1. Introduction

Sulzberger Garbe dermatosis, also known as Oid Oid disease is a variant of eczema first described in 1937 by Dr. Marion B. Sulzberger and Dr. William Garbe. [1] At present not more than a hundred reported cases can be found in literature. Moreover, most of the reports came from North America in the middle of the 20th century. Although the disease has a typical clinical and histopathological picture there is controversy over its existence as an independent disease entity because as of now there are no diagnostic tests specific to the disease. Most reported cases of Oid Oid disease show severe pruritus with discoid exudative plaques associated with lichenification. [2]

2. Case Report

A 40 year old female with itching and skin lesions on the trunk and extremities since 2 months presented to the dermatology OPD. The lesions started on the abdomen, then spread to the back, extremities and face. The lesions which were severely pruritic, initially started as variably sized, erythematous papules and then progressed to violaceous, discoid, faintly exudative lichenoid lesions 2 - 3 cm in size. Based on the classic history and examination a differential of Oid Oid disease was kept and biopsy from the lesions on the arm and the back was sent with the final diagnosis as Sulzberger and Garbe dermatosis.



Violaceous, discoid, lichenoid plaques and erythematous papules present on the abdomen and the legs

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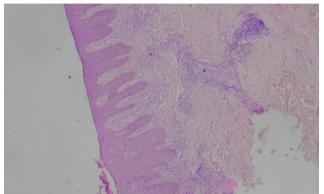
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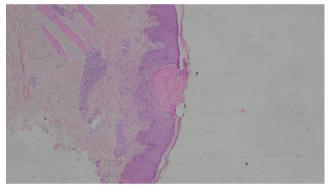
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Histopathological Findings -

The biopsy showed irregular psoriasiform hyperplasia of epidermis with moderate spongiosis. Stratum corneum shows large foci of parakeratosis. Spinous layer is thickened with moderate dyskeratosis. Multiple foci of neutrophilic and eosinophilic micro - abscesses were seen in lower epidermis. Dermis showed moderately dense infiltrate of lymphocytes, plasma cells and eosinophils surrounding thickened blood vessels.



Irregular psoriasiform hyperplasia of epidermis with perivascular chronic inflammatory infiltrate



Micro-abscesses containing neutrophils and eosinophils in epidermis with spongiosis

Lab Findings

The CBC showed moderate eosinophilia (12%) with mild anaemia (11.6) with TLC and platelet count being normal. The rest of the investigations were all within normal limits.

Management and follow up -

The patient was started on oral methylprednisolone 8mg OD which was then slowly tapered along with tab. hydroxyzine and topical mometasone. The lesions resolved within a month along with 80% reduction in itching. The patient was then continued on oral antihistaminics with emollients for another month with no resurgence of the disease.

3. Discussion

In 1979, Sulzberger and Garbe described specific features that are indicative of Oid Oid disease (Table 1) [2] –

- Predilection for the disease among middle-aged males (especially of Jewish origin)
- Before establishing the proper diagnosis, other diseases are frequently considered in the diagnostic process
- The skin lesions typically evolve from elevated discoid, exudative to lichenoid ones and vice versa
- Lesions are fairly common on the genitals (especially on penis in males)
- Small, round or oval, oedematous exanthems that imitate urticaria may occur in the phase of spontaneous regression
- 6) Eosinophilia may occur in the peripheral blood
- Mild endothelium swelling and perivascular infiltrations that consist mainly of lymphocytes, granulocytes, eosinophiles and plasmatic cells are frequently observed in the histopathological examination

The case discussed in this study is of a middle - aged female who is a Hindu by religion which is in contrast to the characteristics of Oid Oid disease described. However, the original disease was described by Sulzberger and Garbe according to the patients seen in their clinic in New York in the 1930's to 1950's which may have led to the data being influenced by the population statistics. [3] There has been a record of this disease in one Indian patient so far excluding our case. [4]

The patient had typical discoid exudative lesions that then evolved into lichenoid lesions and were also associated with intense pruritus. On histopathology parakeratoses and spongiosis was seen along with multiple foci of neutrophilic and eosinophilic micro - abscesses in the lower epidermis. Also, the dermis showed a dense infiltrate of lymphocytes, plasma cells and numerous eosinophils which is in line with the features described by Sulzberger and Garbe and later also by Wilbert Sachs and Neville Kirsch. They also suggested that there is sufficient evidence in the microscopic picture to establish a diagnosis of exudative discoid and lichenoid dermatosis. [5] [6]

The diagnosis of Oid Oid disease can be established once the other, more common dermatoses have been excluded. These include mycoses fungoides and other cutaneous lymphomas, allergic contact dermatitis, Duhring's disease, atopic dermatitis, lichen planus, nummular eczema and prurigo nodularis. [1]

The patient also had significant peripheral blood eosinophilia. She showed a good response to oral corticosteroids with the lesions regressing within a month. Usually, the patients having Oid Oid disease respond well to treatment with oral corticosteroids. Also, there are records of 2 cases of Sulzberger Garbe dermatosis who didn't respond well to corticosteroids and showed a satisfactory response to Azathioprine. [7]

4. Conclusion

Sulzberger and Garbe dermatosis, although described initially in the 1950's has been shrouded in controversy over its existence as a separate disease entity. Also, most of the cases that there initially described were clustered in the Jewish population of North America with the disease being very rarely reported in patients of other ethnicities in other parts of

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the world. Owing to its rarity, no specific diagnostic tests have been established for the same which further makes this a diagnostic conundrum. There was a case of suspected Oid Oid disease in a 7 year old girl where it ultimately concluded that there are no specific clinical or histopathological features that would make it possible to differentiate between Sulzberger and Garbe dermatosis and nummular eczema. [8] Hence, Oid Oid disease is considered as a diagnosis of exclusion in patients who present with a specific clinical and histological picture.

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