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Case Report

Non langerhans cell histiocytoses in genitalia: A rare entity

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Abstract A 35 year old man presented with multiple reddish to skin colored scrotal swellings for the last 8 to 10 months. On biopsy, well circumscribed dense nodular infiltrate composed of histiocytes, lymphocytes and stray plasma cells. On IHC: CD 68 +ve, but CD1a, S100, Langerin, BRAFV600E, CD30 were negative. Based on HPE and IHC reports, a diagnosis of non-Langerhans cell histiocytosis was made. Initially the lesions were treated with intra-lesional corticosteroids, without any significant improvement. As a newer therapy, we tried oral cyclophosphamide 50 mg BD for 2 months, followed by 50 mg OD as maintenance dose, no new lesions were noted.

Key words
Non-Langerhans cell; Cyclophosphamide; Immunohistochemistry.

Introduction

Histiocytoses are disorders which involve monocyte/ macrophage system. Multiple attempts were made for classification of this clinically and histopathological variant group of disorders and syndromes, but no nomenclature was totally acceptable.[1,2] The commonest and the most studied histiocytic disorders are the Langerhans cell histiocytosis (LCH) and the hemophagocytic lymphohistiocytosis (HLH). The Langerhans cell histiocytosis has characteristic features which include extensive growth of histiocytes that bear cytological markers CD1a and langerin (CD 207), that are not present in non-LCH histiocytoses. Practically, other entities belonging to the histiocytoses have nomenclature of "non-Langerhans cell histiocytoses" (non-LCH) together, and as they are rare in occurrence, they are termed as "rare histiocytoses".[2]

NLCDs are a group of disorders where histiocytes aggregate in various tissues. The originating cells of NLCD are macrophage/ monocyte, a cell lineage having wide range of differentiating potential. No details are known about the various pathways of differentiation of monocyte/ macrophages lineage, than any other hematopoietic cells, like lymphocytes and neutrophils. The monocyte/ macrophage may differentiate into any cell types, thus explaining the varied histological features of NLCDs:

1. Resident tissue histiocyte.
2. Foam cell.
3. Foreign body giant cell.
4. Langerhans giant cell.
5. Touton giant cell.
6. Oncocytic histiocyte.[3]

Case report

A 35-year-old male who presented with multiple reddish to skin colored scrotal swellings, present over the past 8 to 10 months. They were

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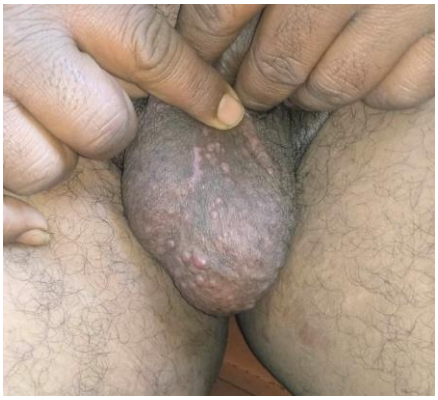

Figure 1

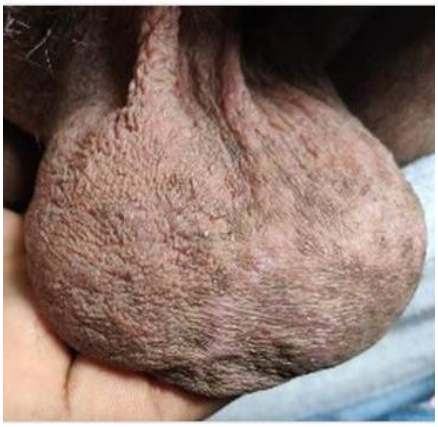

Figure 2

and spread to involve the entire scrotal skin **(Figures 1,2)**. Mild pain and discomfort was present, specially while walking. There was a history of three episodes of similar swellings in the last 10 years.

On Examination it was found that they are multiple, round, reddish to skin coloured papules and nodules ranging between 0.5-1cm in size, mildly tender, smooth and firm on palpation. Area of hyperpigmentation was present. No lymphadenopathy was found. Examination of hair, nail and mucosa was normal. Systemic examination was within normal limits.

Nodular scabies, pseudo lymphoma, sarcoidosis, idiopathic scrotal calcinosis, foreign body granuloma were kept as differential diagnosis.

On biopsy, well circumscribed dense nodular

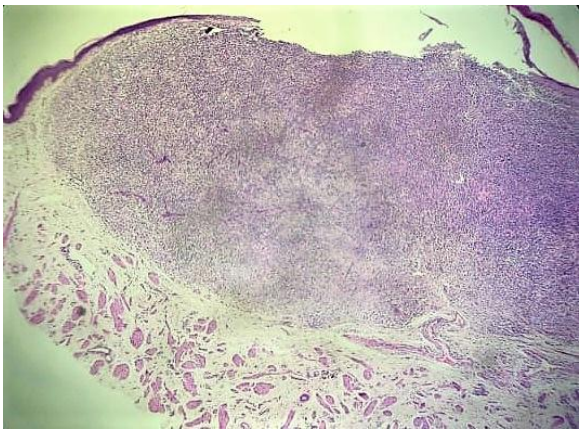

Figure 3

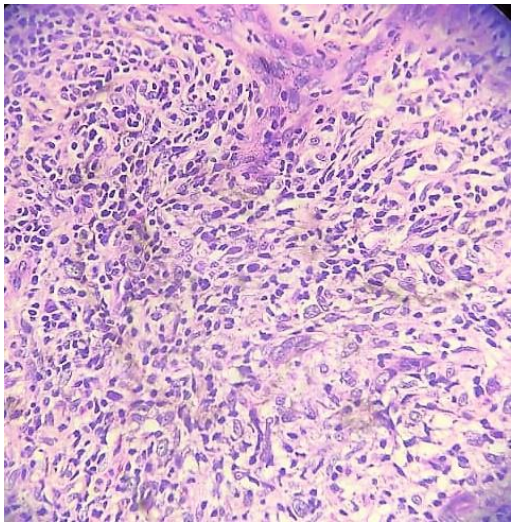

Figure 4

infiltrate composed of histiocytes, lymphocytes and plasma cells were found. On IHC: CD 68 +ve, but CD1a, S100, Langerin, BRAFV600E, CD30 were negative special stains: AFB, PAS, GMS were negative blood parameters-within normal limits **(Figures 3,4)**.

Others differentials were excluded as the patient did not have any itching (nodular scabies and pseudolymphoma excluded). The lesions were firm and not hard like calcinosis. Histopathologically no granuloma either sarcoidal or foreign body granuloma was found. Calcium deposits were not present. Based on HPE and IHC reports, a diagnosis of non-Langerhans cell histiocytosis was made.

Initially the lesions were treated with intra-lesional corticosteroids, without any significant improvement. The patient also did not respond to other immunosuppressives, such as systemic steroids, azathioprine, methotrexate and cyclosporine. As a newer therapy, we tried oral cyclophosphamide 50mg BD for 2 months, followed by 50mg OD as maintenance dose, no new lesions were noted.

Till date, 38 cases of genital LCH have been reported in literature. One case of indeterminate cell histiocytosis has been reported on glans penis, but not a single case of non-LCH over genitalia has been found yet. Hence our case is very unique from both the clinical and management perspective.

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Author's contribution

SC, SK: Identification and management of the case, critical review, final approval of the version to be published.

DD: Management of the case, manuscript writing, final approval of the version to be published.

PS, PSN: Diagnosis of the case, manuscript writing, final approval of the version to be published.

RCG: Management of the case, critical review, final approval of the version to be published.

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