

Peri-ocular, papulo-nodular plaques over bilateral eyelids

A 33-year-old woman presented with an insidious onset, gradually progressive, mildly pruritic, multiple, raised lesions over her bilateral eyelids for one year. She did not have any systemic disease and her primary concern was aesthetic. Cutaneous examination revealed multiple discrete to coalescent, yellow to reddish-brown, papulo-nodular plaques present symmetrically and circumferentially over bilateral periocular area [Figure 1]. There was no involvement of other sites such as the flexures and mucosae. Haematological examination including lipid profile was within normal limits. A biopsy from the lesions showed dense, spindle cell proliferation arranged in the form of short fascicles and focal storiform pattern in the dermis [Figure 2]. On high-power

magnification, mononuclear histiocytic cell proliferation was seen in the dermis [Figure 3]. Immunohistochemistry (IHC) showed positive staining for vimentin and CD-68 and negative staining for CD-34, smooth muscle actin, S-100, desmin, myogenin and beta-catenin [Figures 4a–4d]. A bone marrow biopsy was done, which revealed a hypercellular marrow with myeloid prominence with leukocytosis and eosinophilia. JAK-2 Exon 14 mutation assay revealed no abnormality. A haematology consult was taken to rule out systemic involvement.

Question

What is your diagnosis?



Figure 1: Multiple yellow to red-brown papulo-nodular plaques over peri ocular area.

How to cite this article: Sandhu J, Singh A, Gupta SK, Singh A, Garg B. Peri-ocular, papulo-nodular plaques over bilateral eyelids. *Indian J Dermatol Venereol Leprol.* 2024;90:385–7. doi: 10.25259/IJDVL_377_2023

Received: April, 2023 **Accepted:** August, 2023 **Epub Ahead of Print:** January, 2024 **Published:** April, 2024

DOI: 10.25259/IJDVL_377_2023 **PMID:** 38314972

This is an open-access article distributed under the terms of the Creative Commons Attribution-Non Commercial-Share Alike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

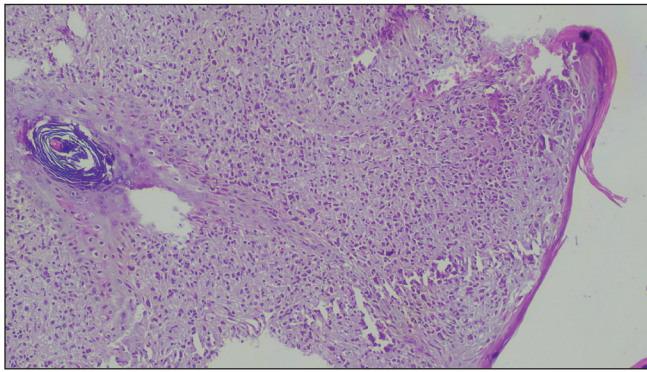


Figure 2: Diffuse infiltration by histiocytes exhibiting focal storiform pattern (Haematoxylin and Eosin, 100x).

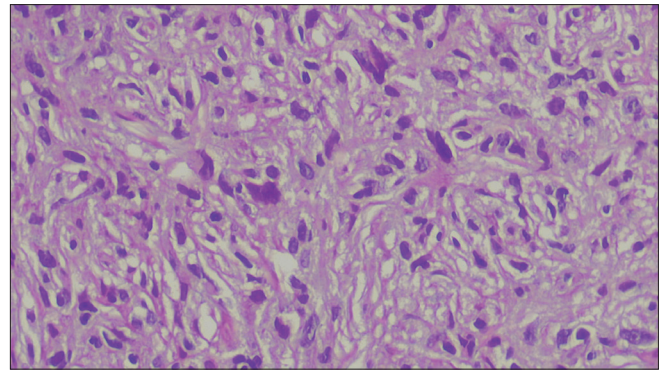


Figure 3: Mononuclear histiocytic cell proliferation seen in the dermis (Haematoxylin and Eosin, 400x).

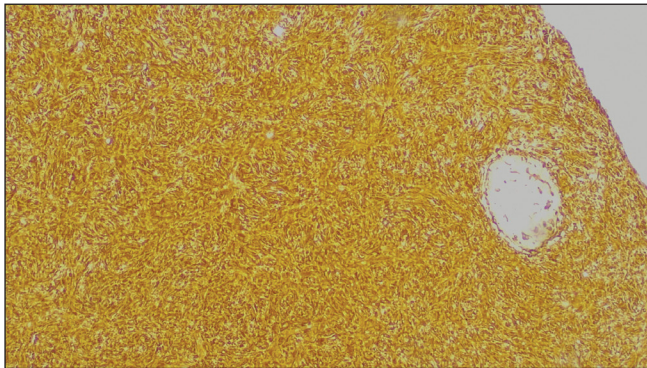


Figure 4a: Immunohistochemistry (IHC) (100x): Positivity of vimentin.

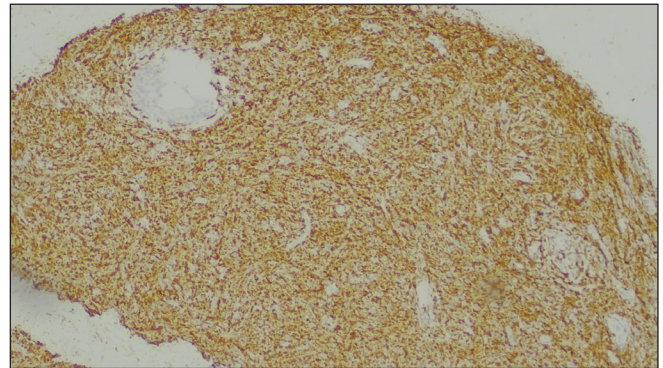


Figure 4b: Immunohistochemistry (IHC) (100x): Positivity for CD-68.

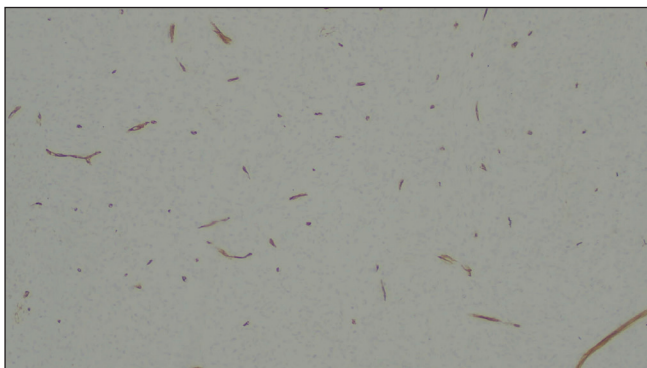


Figure 4c: Immunohistochemistry (IHC) (100x): Negative IHC for CD-34.

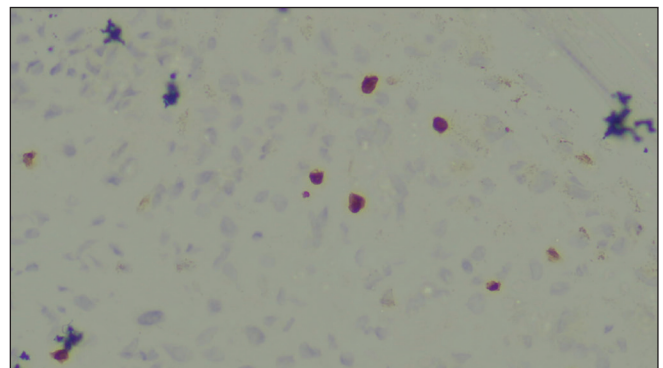


Figure 4d: Immunohistochemistry (IHC) (100x): Negative IHC for Ki-67.

Answer

Diagnosis: Progressive nodular histiocytosis

Discussion

Progressive nodular histiocytosis (PNH) is a rare variant of non-Langerhans cell histiocytosis affecting the skin and mucous membranes. It was first described by Taunton *et al.* in 1978.¹ PNH presents as persistent, progressive, disseminated papulo-nodules involving head, neck and trunk.² Severe facial involvement in some cases may lead to leonine facies, and ophthalmological involvement may occur.² Present case was an unusual case of progressive nodular histiocytosis without dissemination and localised to the eyelids. Skin biopsy was performed with the differential diagnosis of necrobiotic xanthogranuloma (NXG) and lipoid proteinosis. However, on the basis of clinical, histopathological examination and IHC, we arrived at a diagnosis of progressive nodular histiocytosis.

To the best of our knowledge, less than 25 cases of progressive nodular histiocytosis (PNH) have been reported so far. It is usually characterised by its progressive course and multiple disseminated, typical pedunculated nodules. Our case is unusual because the patient did not have disseminated lesions. The histopathology and IHC, however, were consistent with the diagnosis of PNH. A possible association with chronic myeloid leukaemia has been reported with long-standing disease.³ Therefore, JAK-2 Exon-14 mutation assay was performed to rule out myeloproliferative association. However, more data is needed to substantiate this association.

PNH should be differentiated from other types of histiocytosis by its distribution, morphology and systemic involvement. It has a prominent facial distribution with a progressive but benign course as compared to juvenile xanthogranuloma and benign cephalic histiocytosis which usually shows spontaneous regression.³ Xanthoma disseminatum is commonly associated with diabetes insipidus and respiratory tract involvement, which is not seen in PNH. Generalised eruptive histiocytosis has a generalised and symmetrical distribution, which is usually benign and may show spontaneous resolution.² There are reports with lesions predominantly involving head and neck but disseminated lesions were also present in these patients.^{2,4-7}

This case is of particular interest due to its unusual clinical presentation as well as lack of dissemination. PNH is a rare and has esoteric cutaneous pathology, which may pose a diagnostic challenge to the dermatologists.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

**Jaspriya Sandhu, Amandeep Singh,
Sunil Kumar Gupta, Aminder Singh¹, Bhavna Garg¹**

Departments of Dermatology, Venereology & Leprology and ¹Pathology,
Dayanand Medical College & Hospital, Ludhiana, Punjab, India

Corresponding author:

Dr. Jaspriya Sandhu,
Departments of Dermatology, Venereology & Leprology,
Dayanand Medical College & Hospital,
Ludhiana, Punjab, India.
sandhu.jaspriya@gmail.com

References

1. Taunton OD, Yeshurun D, Jarratt M. Progressive nodular histiocytoma. *Arch Dermatol* 1978;114:1505–8.
2. Nofal A, Assaf M, Tawfik A, Elsayed N, Nofal E, Elnakib N, *et al.* Progressive nodular histiocytosis: A case report and literature review. *Indian J Dermatol* 2011;50:1546–51.
3. Gonzalez Ruiz A, Bernal Ruiz AI, Aragonese Fraile H, Peral Martinez I, Garcia Muñoz M. Progressive nodular histiocytosis accompanied by systemic disorders. *Br J Dermatol* 2000;143:628–31.
4. Numbere N, Pukhalskaya T, Bowman B, Campbell K, Smoller B. Progressive nodular histiocytosis: Report of a case and review of the literature. *Case Rep Pathol* 2021;2021:5531820.
5. Williams A, Thomas AG, Kwatra KS, Jain K. Progressive nodular histiocytosis associated with Eale's disease. *Indian J Dermatol* 2015;60:388–90.
6. Roldan MS, Choc C, Mansilla JJ, Riley G. Progressive nodular histiocytosis: An unusual disorder. *Dermatol Online J* 2021;27:13030/qt4t37r77d.
7. Chuang FC, Chern E, Wu WM. Progressive nodular histiocytosis: A rare type of xanthogranuloma. *Dermatol Sin* 2011;29:98–100.