Photo Essay

Disfiguring periocular yellow nodules in a normolipemic patient

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Key words: Histiocytosis, nodules, periorbital

A 35-year-old female presented with 1-year duration of numerous asymptomatic, firm, yellowish-brown nodules and plaques involving both eyelids including the tear trough [Fig. 1a]. A single yellowish nodule was also present over the elbow [Fig. 1b]. There was no history of ulceration, diminution of vision, voice change, polyuria, or other systemic symptoms. Sclera and conjunctiva were not involved and further ocular examination did not reveal any abnormality. Rest of the cutaneous examination including oral and nasal mucosae was found to be normal. Fasting lipid profile, serum protein electrophoresis, urine and plasma osmolarity, and thyroid function tests were normal. Biopsy done from the lid nodule showed a dermal tumor composed of sheets of spindle-shaped foamy histiocytes [Fig. 2a and b]. The histiocytes stained positive for CD68 and negative for S100 [Fig. 2c]. Touton giant cells were not seen. A diagnosis of progressive nodular histiocytosis (PNH) was thus made. Patient was given a trial of imatinib 400 mg daily for 4 months. Due to inadequate response, the option of surgical excision was offered, but the patient did not follow-up.

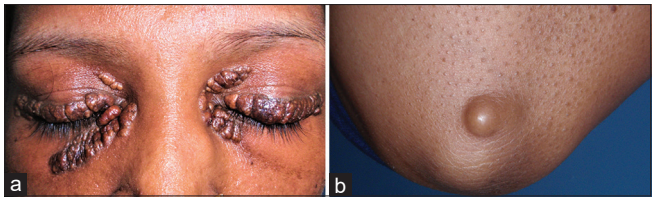

Figure 1: (a) Multiple yellowish-brown papules coalescing to form plaques involving both eyelids and orbital margins. (b) Single yellowish firm nodule over the elbow

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Discussion

PNH, a variant in the spectrum of non-Langerhans cell histiocytosis (NLCH), is characterized by multiple disfiguring yellowish-brown nodules.[1] It is seen in adults with a predilection for the face and progressive course spanning decades. In most cases, the disease is purely cutaneous with the absence of systemic symptoms. However, there are occasional reports of PNH associated with myeloid leukemia, precocious puberty, dyslipidemia, hypothyroidism, and growth hormone deficiency. Oropharyngeal and laryngeal involvement can lead to dyspnea and dysphagia.[2] Ocular involvement in the form of epibulbar infiltrates[2] and association with Eale's disease has been reported.[3] Chemotherapy with corticosteroids, methotrexate, imatinib, and vinblastin has been tried with variable success and the disease is often treatment resistant. Disfiguring lesions can be surgically excised but recurrences are not uncommon.[4]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

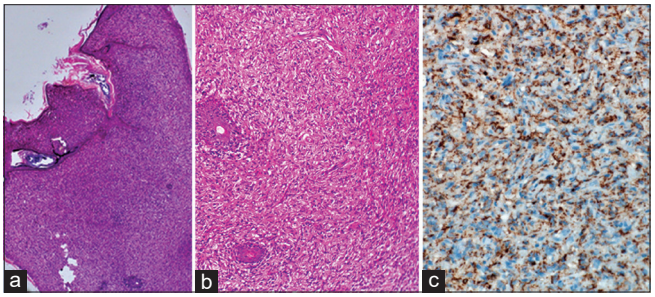

Figure 2: (a) Compact dermal tumor with loss of grenz zone. (Hematoxylin and Eosin, 10×). (b) Sheets of spindle-shaped histiocytes with foamy cytoplasm seen in the dermis. (Hematoxylin and Eosin, 40×). (c) Histiocytes stained positive for CD68 and negative for S100 (Immunohistochemistry for CD68, 100×)

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Conflicts of interest

There are no conflicts of interest.

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