Multifocal Cutaneous Sarcoidosis Presenting as Lid Ectropion

Sir,

Sarcoidosis is an idiopathic multisystem granulomatous disorder that most commonly affects the lungs. Cutaneous involvement is seen in 30% cases and is rightly termed as the ‘great imitator’ due to varied morphological presentations. Identification of non-caseating granulomas and exclusion of other causes of granulomatous dermatoses is diagnostic. However, systemic involvement is looked for in all cases of cutaneous sarcoidosis. Periocular adnexal sarcoid is not very commonly seen and can masquerade as various other dermatoses. Herein, we report a case of multifocal cutaneous sarcoidosis presenting as lid ectropion.

A male in his forties presented with 4 years history of multiple yellowish asymptomatic plaques over both the upper eyelids and neck. Lesions over the left eyelid resulted in thickening of the lid and ectropion [Figure 1a and b]. He gave history of a similar lesion over the right shoulder which had resolved, spontaneously. He denied any systemic complaints and rest of the mucocutaneous examination was normal. Dermatoscopy of eyelid papules and neck revealed yellowish orange areas with dilated vessels suggesting a granulomatous pathology. Ophthalmological examination showed restriction of upper eyelid movements, loss of cilia and left eyelid ectropion. There was no evidence of uveitis or dacryoadenitis. Biopsy done from the plaque on the neck, showed naked granulomas consisting of epithelioid cells in the dermis extending up to the subcutis [Figure 2a and b]. Staining with reticulin stain was intact and Ziehl Neelsen stain did not reveal any acid-fast bacilli. Serum angiotensin converting enzyme levels, serum calcium and chest radiographs were normal. FDG-PET (fluorodeoxyglucose positron emission tomography) scan showed FDG avid areas involving bilateral eyelids without orbital involvement, anterior neck and right shoulder in the cutaneous and subcutaneous plane corresponding to the site of lesions. With the diagnosis of cutaneous sarcoidosis, the patient was initiated on oral prednisolone (0.5 mg/kg) and hydroxychloroquine 300 mg once daily and he had a dramatic improvement in skin lesions, lid infiltration and ectropion within 1 month of treatment [Figure 3a and b]. Patient was satisfied with the result and no reconstructive surgery was planned for the mild residual ectropion. He was advised regular follow-up so as to pick up early recurrence.

Ocular sarcoidosis is fairly common (25–60%) with anterior uveitis being the most common presenting feature. Orbital involvement in sarcoidosis is rare with only 11.5–17% involving the eyelids. Various presentations include “millet seed” nodules, ulcerated nodules, plaques, and peri-orbital swelling. Full thickness scarring of anterior and posterior lamella can result in ectropion or entropion, symblepharon, and lid notching. Topical and intralesional corticosteroids are the first line therapy for lesions limited to the skin. Systemic corticosteroids remain the treatment of choice for rapidly progressive, generalized, or highly disfiguring skin disease. Although the index case did not have any evidence of systemic involvement, systemic steroids were initiated in view of the disfiguring lid lesions. Antimalarials like hydroxychloroquine are effective in preventing the development of new skin lesions. In recalcitrant disease, steroid sparing agents like methotrexate can also be used. In previous reports of sarcoidosis leading to distortion of lid architecture, patients had to undergo reconstructive surgery and wedge resection to prevent trichiasis and corneal scarring.

Figure 1: (a) Multiple discrete skin coloured, yellowish brown papules coalescing to form plaques in the periorbital area associated with thickening and ectropion of the left eyelid. (b) Yellowish plaque seen over the anterior aspect of the neck.

Figure 2: (a) Biopsy from the plaque on the neck- Epidermis is mildly thinned out. Whole of dermis shows nodular collections of epithelioid cell granulomas (Hematoxylin and eosin 20x). (b) The granulomas are naked and do not show any lymphocytic cuffing or admixture of lymphocytes (Hematoxylin and eosin 400x).

Figure 3: (a) Significant improvement in periorbital lesions including lid ectropion after 1 month of treatment. (b) Near complete resolution of the plaque over the neck.
We want to highlight the importance of early diagnosis and initiation of systemic steroids regardless of presence of systemic involvement in cases with disfiguring lesions. This could prevent permanent distortion of lid architecture and prevent surgical intervention.

**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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**Conflicts of interest**

There are no conflicts of interest.

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