Uncommon presentations of lupus miliaris disseminatus faciei

Sir,

Lupus miliaris disseminatus faciei is an uncommon granulomatous disorder predominantly involving the centro-facial area, including the region around the eyelids. It usually presents as multiple skin colored or yellowish brown papules and pustules, which heal to leave behind pock-like scars. Although extrafacial involvement and a nodular morphology of lesions have been reported, these are very uncommon.[1-3] Herein, we report four patients of lupus miliaris disseminatus faciei with simultaneous involvement of facial and extra-facial regions. Three of the patients also had prominent nodular and a few nodulo-cystic lesions. We were unable to find previous reports of this presentation.

A 32-year-old male patient presented with multiple, disfiguring "acne-like" lesions on the face, chest and upper back since 6 months. The lesions were asymptomatic but were gradually increasing in extent with appearance of new papules on face. During this time, none of the lesions showed spontaneous regression. On examination, multiple, brownish papular lesions with central scarring were noted on the forehead, cheeks and helix of the ears [Figure 1a]. Lesions of similar morphology were also present on the chest and upper back. There were multiple, firm, freely mobile, nodulo-cystic lesions on the forehead [Figure 1a and b]. Chest radiography was normal and Mantoux intradermal test was negative. A skin biopsy obtained from a papular lesion showed multiple epithelioid cell granulomas in the periappendageal dermis [Figure 2a and b]. A slit-skin smear for lepra bacilli obtained from the papules was negative. Histopathological examination of a nodulo-cystic lesion showed a cyst wall made of fibro-collagenous tissue, devoid of lining epithelium (pseudo-cyst). Multiple compact epithelioid cell granulomas with giant cells were also noted in the cyst wall [Figure 2c and d]. Based on these features, a clinical diagnosis of lupus miliaris disseminatus faciei was made and the patient was treated with oral minocycline, 100 mg twice daily. After 18 months of follow-up, there was complete resolution of nodulo-cystic lesions. Most of the papular lesions had healed with pock- like scars [Figure 1a and b].

Figure 1: Multiple brownish papules on the cheeks, forehead and concha with a prominent nodule on forehead (arrow) (a). Following treatment with oral minocycline, there was a complete healing of the lesions with pock like scars (b)

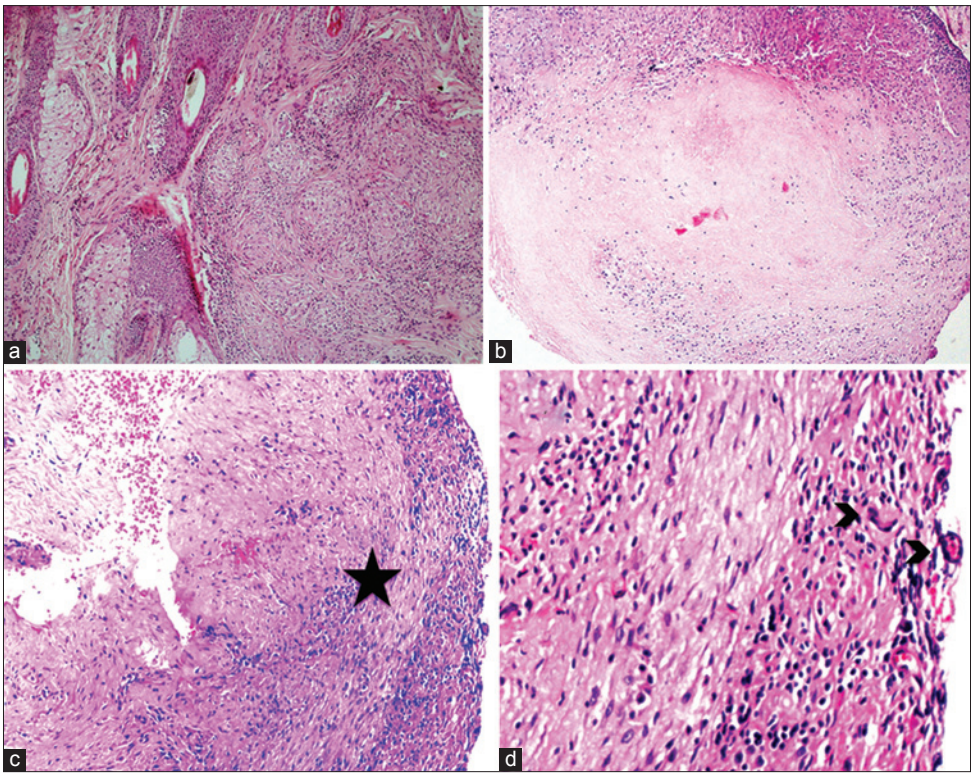

Figure 2: Compact non-necrotizing granulomas around hair follicles (a) and large area of caseous necrosis bordered by epithelioid histiocytes, H and E, ×200 (b). Cystic lesion showing a fibrous capsule devoid of epithelium (star, H and E, ×200) (c). The cyst wall contains many epithelioid cells and giant cells (arrow, H and E ×400) (d)

Following this, we came across three other patients of lupus miliaris disseminatus faciei with extra-facial lesions, two of them having prominent nodulo-cystic lesions (patients 2 and 3). Similar to our index case, all three patients had skin colored to reddish-brown papular lesions distributed predominantly on the centro-facial region. All of them had extra-facial lesions involving the neck and upper chest. Patient 2 also had involvement of axilla and scalp resulting in scarring alopecia [Figure 3a and b]. Chest radiography was normal and Mantoux intradermal test was negative in all three cases. The salient investigations, histopathological findings and treatment response are summarized in Table 1.

The nosological status of lupus miliaris disseminatus faciei has been a matter of debate ever since its first description. Radcliffe-Crocker initially coined the term 'acne agminata' to describe the condition although it bore only a remote resemblance to acne and the lesions were not always grouped.[4] The other names of this entity, that is, lupus disseminata and granulomatous rosacea have their detractors. More recently, a broad term "facial idiopathic granulomas with regressive evolution" (FIGURE)

Figure 3: Patient 2 showing reddish brown papular lesions over the scalp causing scarring alopecia (a) reddish brown papular lesions on the face and nodular lesions on the neck (b). Reddish brown papulo-nodular lesions involving the axilla (c)

Table 1: Clinical and histological features and treatment response

Patient	Age (years)/ sex	Disease duration (months)	Clinical features		Differential diagnosis	Histopathology	Treatment	Response
			Site	Lesional morphology				
1	32/male	6	Forehead, cheeks, chest and upper back	Brownish papular lesions with central scarring	Acne cyst	Multiple epitheloid cell granulomas in peri-appendageal location. Stain for lepra bacilli was negative	Minocycline 100 mg once daily × 5 months	Complete response
			Forehead	Multiple, firm, freely mobile, nodulo-cystic lesions				
2	36/male	7	Face and scalp	Reddish-brown papular lesions resulting in scarring alopecia on scalp. Some of these lesions had healed to leave behind pock like scars	Papular sarcoidosis	Perivascular and peri-appendageal lympho-histiocytic infiltrate with multiple epithelioid cell granulomas	Doxycycline 100 mg twice a day × 12 weeks	No response
			Neck and axilla	Multiple nodular and nodulo-cystic lesions				
3	25/male	6	Eyelids, preauricular region and neck	Reddish-brown papules, plaques and nodulo-cystic lesions with central scarring	Acne cyst	Multiple compact granulomas in deep dermis in perivascular and peri-appendageal location with areas of necrosis	Doxycycline 100 mg twice a day	Lost to follow-up
4	18/male	2	Cheeks, peri-ocular area and neck	Skin colored and brownish papules	Sarcoidosis, granulomatous rosacea	Perivascular and peri-appendageal lymphohistiocytic infiltrate forming compact granulomas with small areas of necrosis	Doxycycline 100 mg twice a day × 12 weeks	Significant improvement

has been proposed, which would unify this entity along with other non-specific granulomatous disorders occurring on centro-facial area under a single name.[5]

Classically, the lesions in lupus miliaris disseminatus faciei are papular or pustular and are confined to the face. All four patients in the present series had lesions extending beyond the facial region. Another atypical feature seen in the patients was the presence of nodular and nodulo-cystic lesions. Nath *et al*. described a patient of lupus miliaris disseminatus faciei with reddish-brown papules and nodules on facial and extra-facial sites.[3] Their patient resembled our cases although the lesions were less inflammatory and not cystic. Such nodulo-cystic lesions need to be differentiated from epidermoid cysts and acne cysts. Sanz-Sánchez *et al*. reported a patient of lupus miliaris disseminatus faciei with co-existing epidermoid cyst and hypothesised that the rupture of the cysts may be related to the appearance of lupus miliaris disseminatus faciei.[6] Rupture of acne nodules and epidermoid cysts can incite a granulomatous response which can closely mimic lupus miliaris disseminatus faciei.[7] However, in such cases, the inflammation is generally suppurative, associated with rupture, distortion and destruction of appendages ultimately leading to scarring.[7] Caseous necrosis, typical of lupus miliaris disseminatus faciei is not seen in ruptured cysts.

Hillen *et al*. and Bedlow *et al*. have each reported a case of axillary acne agminata.[1,2] Similar to our third patient, the case reported by Hillen *et al*.[1] also had classical lesions of lupus miliaris disseminatus faciei on the face. Interestingly, our patient additionally had scalp involvement resulting in scarring alopecia. We were able to find only one other such published report.[8]

Nodular lesions in lupus miliaris disseminatus faciei are under-recognized and/or under-reported.

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Conflicts of interest

There are no conflicts of interest.

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