Keratosis lichenoides chronica showing significant response to acitretin

Sir,

Keratosis lichenoides chronica or Nekam's disease is a rare disorder presenting with lichenoid papules arranged linearly or in a reticulate pattern over the extremities along with facial erythema in seborrheic distribution and oral erosions.

We report a 24-year-old woman, who presented with asymptomatic keratotic papules and plaques over the face, trunk, upper extremities, gluteal area, thighs and legs for the past seven years. The lesions began as erythematous papules and plaques over the centrofacial area which later developed into the firm, non-tender keratotic papules and plaques, followed by progression to other body parts. She also complained of erythema and scaling in the seborrheic areas accompanied by photosensitivity. At presentation, she had a symmetrical eruption on the face, trunk and extremities comprised of well-defined, non-tender, thick keratotic papules and plaques of size 0.5–3 cm with overlying adherent thick yellowish-brown scale [Figures 1a-c]. The papules were arranged linearly at places. Erythematous, semi adherent, mildly scaly plaques were present over the nasolabial folds and forehead. There was a single painless erosion over the hard palate with mild conjunctival erythema. Nails did not show any abnormality. Based on the clinical morphology we made a clinical diagnosis of Nekam's disease with differential diagnosis of hypertrophic lichen planus and verrucous psoriasis.

Biopsy from a keratotic papule on the back showed compact hyperkeratosis, papillomatosis, irregular acanthosis with a dense band-like infiltrate of lymphocytes and histiocytes in upper dermis impinging on the overlying epidermis with focal basal cell damage and occasional necrotic keratinocytes and absent basement membrane thickening [Figures 2a and b]. There was mild to moderate similar infiltrate present around the eccrine glands in the deep dermis without any appreciable mucin in the dermis [Figure 2c]. Overall the histopathological features were suggestive of a hypertrophic lichenoid tissue reaction. Based on the clinical and histopathological findings, the diagnosis of Nekam's disease or keratosis lichenoides chronica was made. Routine biochemical investigations (complete blood count, liver function test, kidney function test, lipid profile and blood sugar) were within normal limits and anti-nuclear antibody and dsDNA were negative. The patient was treated with acitretin, 25 mg orally twice a day, after taking a written consent explaining the teratogenic side-effects of acitretin; along with topical desonide 0.05% cream once a day over the face for facial erythema for two weeks. At the end of six months, there was an 80–90% improvement, with flattening of keratotic papules with post-inflammatory hypopigmentation [Figures 3a and b]. Erythema and scaling in the seborrheic areas and oral ulcers persisted but had diminished.

Keratosis lichenoides chronica is a rare papulosquamous disorder characterized by erythematous to violaceous papules and plaques arranged in a very characteristic linear and reticulate pattern over the extremities. In addition, patients have facial erythema or seborrheic dermatitis along with oral involvement presenting as persistent erosions and ulceration; and ocular involvement presenting as blepharitis and conjunctivitis.[1] Nails may show onychodystrophy and discoloration.[2] The disease is more common in adults and is rare in children. Oral, genital and nail involvement are noted frequently in adults while alopecia and pruritus are frequent in children.[3] Histopathology shows a lichenoid tissue reaction characterized by epidermal atrophy or acanthosis, parakeratosis, follicular plugging and lichenoid lymphocytic infiltrate with basal cell vacuolization.[2,4] Unlike lichen planus, the infiltrate contains plasma cells and extends to the deep dermis around eccrine glands.[5] The exact etiopathology of this disease is unknown but few factors like mechanical injury, trauma, autoimmunity, infections, drugs and hematological malignancies have been associated.[6-9]

Although, there is no definitive laboratory test for the diagnosis of keratosis lichenoides chronica, clinical and

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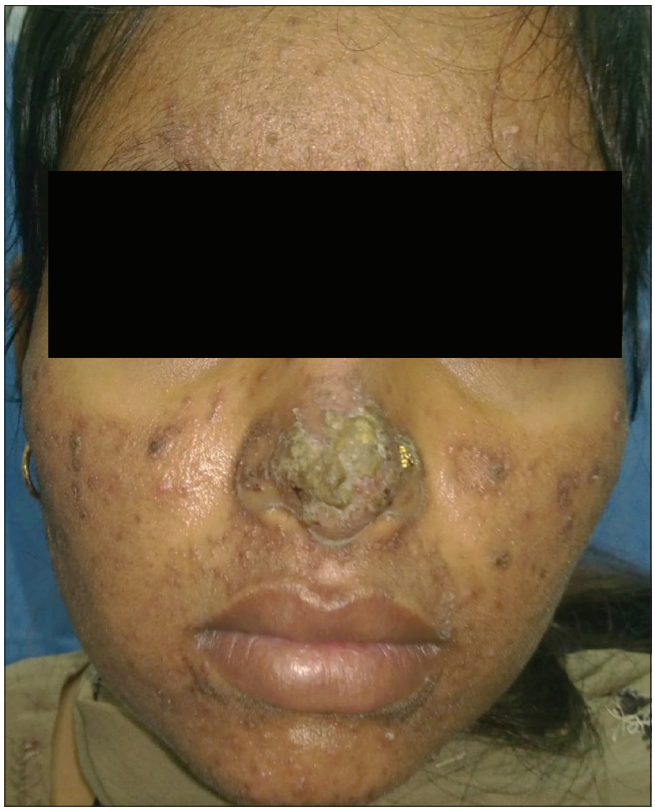

Figure 1a: Multiple hyperkeratotic papules and plaques over the centrofacial region with erythema over the forehead, nose, cheek and perioral region

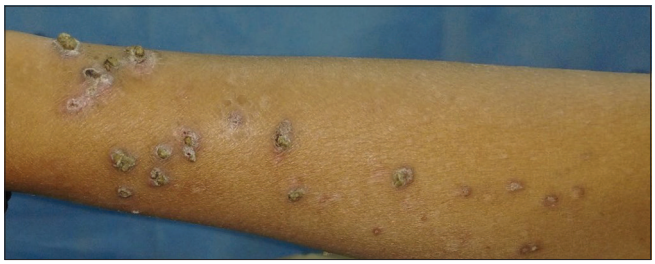

Figure 1b: Hyperkeratotic papules and plaques arranged linearly over the arms

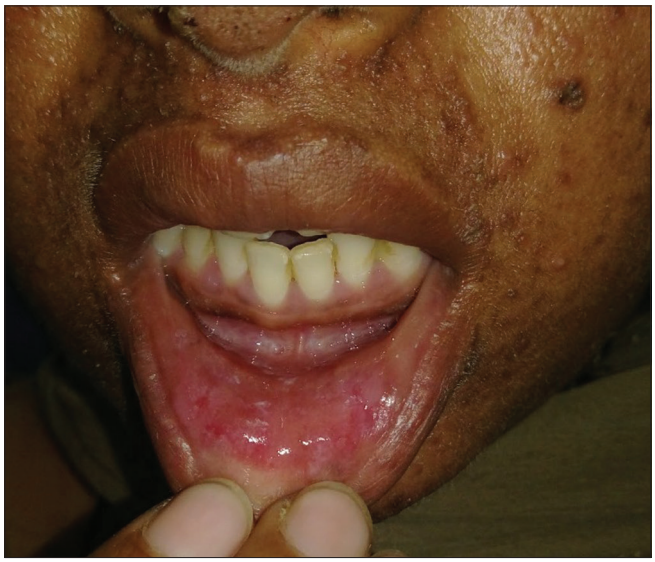

Figure 1c: Superficial erosions over the mucosal aspect of the lower lip

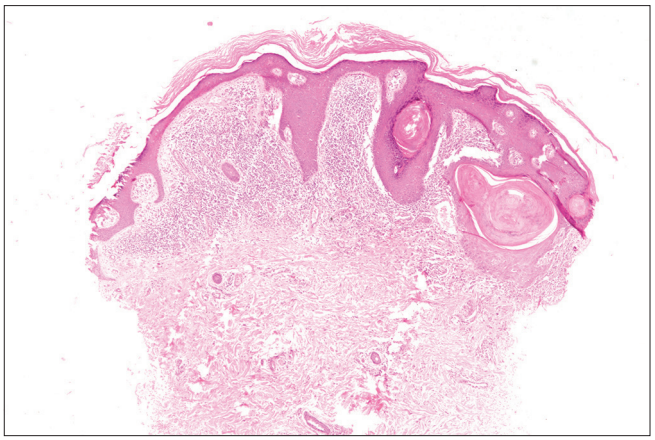

Figure 2a: Histopathology (H and E, ×40) showing hyperkeratosis, papillomatosis, irregular acanthosis, focal basal cell damage with a dense band-like infiltrate of lymphocytes and histiocytes in the upper dermis

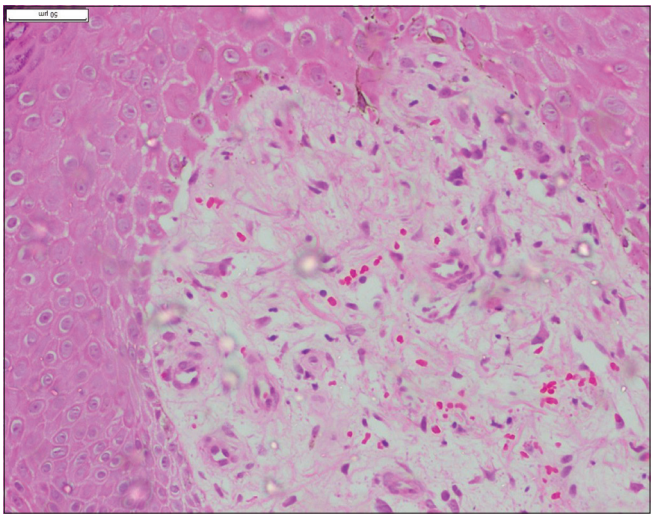

Figure 2b: Histopathology (H and E, ×400) showing vacuolar degeneration of the basal layer of the epidermis

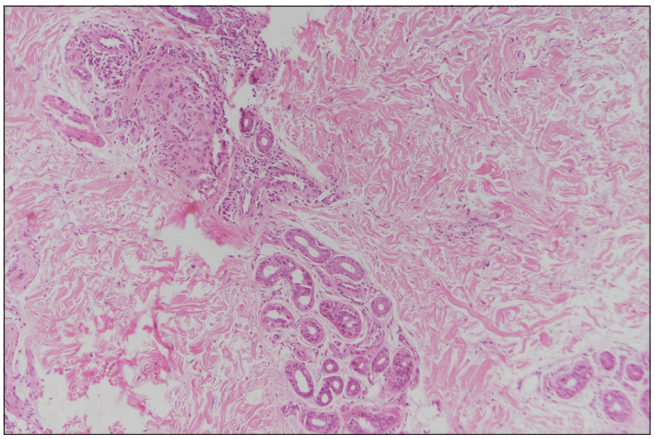

Figure 2c: Histopathology (H and E, ×100) showing deeper lymphohistiocytic infiltrate around the appendages and eccrine glands

histopathological clues are sufficient to make the diagnosis. Treatment is challenging and various treatment options such as oral and systemic steroids, vitamin D analogs, methotrexate,

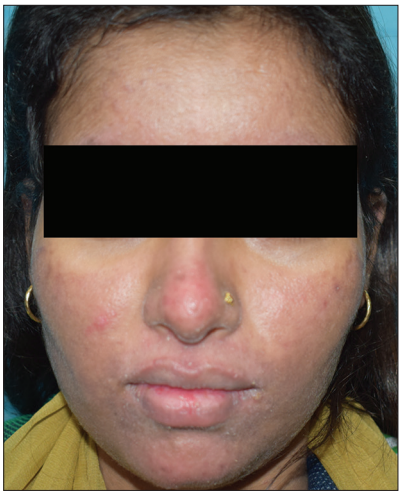

Figure 3a: Posttreatment resolution of the hyperkeratotic papules and plaques over the face with the persistence of facial and conjunctival erythema

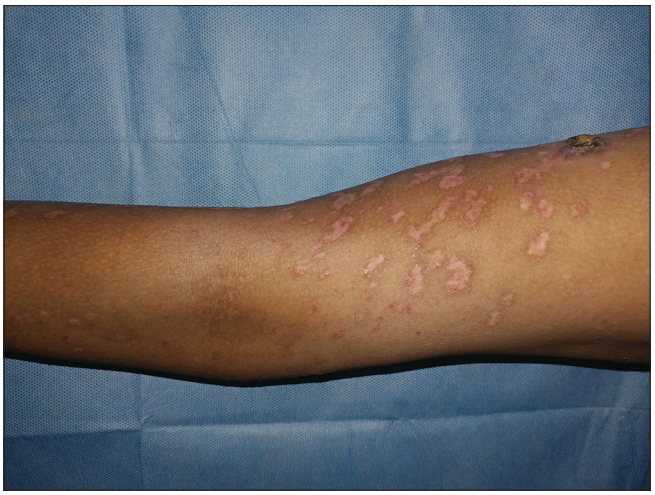

Figure 3b: Posttreatment resolution of the linearly arranged hyperkeratotic papules over the arms

cyclosporine, antimalarials and azathioprine have been tried previously without much improvement. Oral retinoids (acitretin, isotretinoin) were found to be the most effective among all the currently available options.[10,11] Improvement begins in a month and significant improvement can be expected in 4–6 months.[11,12] A review of 30 patients of Nekam's disease treated with retinoids showed a partial response in 6 (20%) and a complete response in 11 (36.6%) patients.[13]

Our patient noticed a significant improvement in the skin lesions with acitretin. A short course of oral steroid was tried for persistent facial erythema without much improvement. Though we were unable to achieve complete remission, the response was sufficient to improve the facial appearance and quality of life of our patient.

Declaration of patient consent

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

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Nil.

Conflicts of interest

There are no conflicts of interest.

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