Olmsted Syndrome with Lateral Supraciliary Madarosis and Clubbing: A Rare Case Report

Abstract

Olmsted syndrome (OS) is a rare congenital, mutilating palmoplantar keratoderma first described by Olmsted in 1927. It starts in the neonatal period or in childhood, and has a slow but progressive disabling course. We report the case of a 16-year-old boy who presented with keratoderma of the palm and soles since childhood with lateral supraciliary madarosis and clubbing. The patient was started on oral retinoids and topical keratolytics and had partial improvement in 2 months. Keratoderma of the palms and soles along with lateral supraciliary madarosis and clubbing in our case is a very rare finding, and to the best of our knowledge, has not been reported so far.

Keywords: *Clubbing, madarosis, Olmsted syndrome, palmoplantar keratoderma*

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Introduction

Olmsted syndrome (OS) is a rare congenital disorder of keratinization characterized by symmetrical sharply defined mutilating palmoplantar keratoderma.[1] Classically, it is characterized by bilateral mutilating palmoplantar keratoderma (PPK) with periorificial keratotic plaques, however, it shows high clinical variability.[2] There is a paucity of literature on this rare syndrome with thin and sparse eyebrows.[3] We report a case of OS who had clubbing and lateral supraciliary madarosis.

Case Report

A 16-year-old boy presented with complaints of thickening of palms and soles since the age of 2 months. He also suffered fromrecurrent episodes of papular lesions with fissuring at the angle of mouth and gradual thinning of lateral eyebrows [Figures 1a and b]. The child was born to nonconsanguineous parents and there was no history of similar disease in the family. On examination, the patient was of short stature with normal physical and mental development. There was no evidence of any associated systemic disease. On cutaneous examination, diffuse bilateral symmetrical, dirty, yellowish, heaped-up, fissured hyperkeratotic plaques were present on both palms and soles [Figure 2]. The keratotic plaques were tender on palpation. Hyperkeratotic scaly plaques were also present on the dorsum of hand [Figure 3]. The fingers and toes showed fixed flexion deformity. The 5th toes were amputated due to constriction bands previously. Clubbing was present in all fingers [Figure 3]. Examination of the scalp hair, teeth, eyes, and joints were normal. Routine laboratory tests and serum zinc level were all within normal range. Slit smear examination for acid fast bacilli was negative.

The patient was prescribed capsule acitretin (0.5 mg/kg/day) along with topical keratolytics (20% urea cream) and emollients. There was partial improvement after 8 weeks and the patient was able to walk [Figure 4].

Discussion

OS was first described in 1927 by Olmsted as a combination of congenital palmoplantar keratoderma, and periorificial hyperkeratosis.[1] A review article by Duchatelet *et al*. reported a total 73 OS cases worldwide until December 1, 2014.[4]

Although autosomal dominant, recessive and X-linked inheritance has been reported, most cases are sporadic. Mutation in *TRPV3* (Transient receptor potential vanilloid-3) on chromosomal region 17p13 has been reported in most OS patients. Mutations in *MBTPS2* (membrane-bound transcription factor protease, site 2) gene

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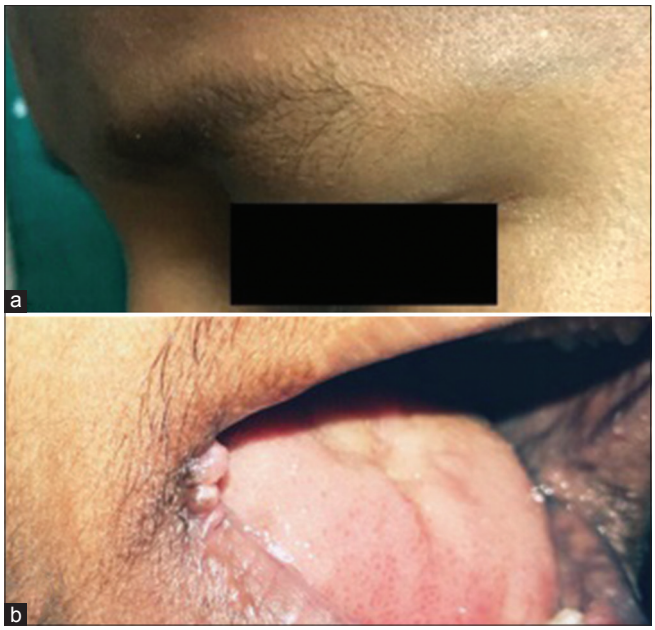

Figure 1: (a) Lateral supraciliary madarosis; (b) periorificial keratotic papules

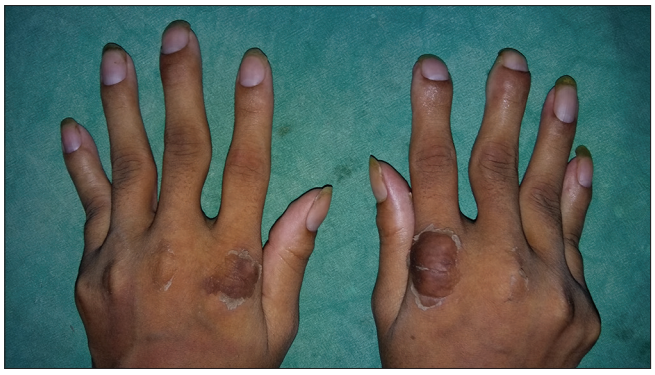

Figure 3: Hyperkeratotic scaly plaques on the dorsum of hands along with clubbing

encoding a zinc metalloprotease has also been reported in recessive X-linked OS.

The disease starts in the neonatal period or childhood and has a slow but progressive and disabling course.[5] Palmoplantar keratoderma may lead to flexion deformity and spontaneous amputation of the fingers.[5] Our patient also had keratoderma on the palms and soles which gradually progressed as painful, massive, hyperkeratotic fissured plaques with flexion deformities of digits and ultimately leading to amputation of bilateral 5th toes.

Most OS patients presented with hair abnormalities including alopecia (diffuse, universal, or patchy), hypotrichosis, sparse, thinning, curly, woolly, coarse, dry, or easily broken hair.[4] Madarosis had also been reported in earlier few studies.[3] Though our patient had no scalp hair abnormalities on dermoscopy and microscopic hair examination, he had supraciliary lateral madarosis.

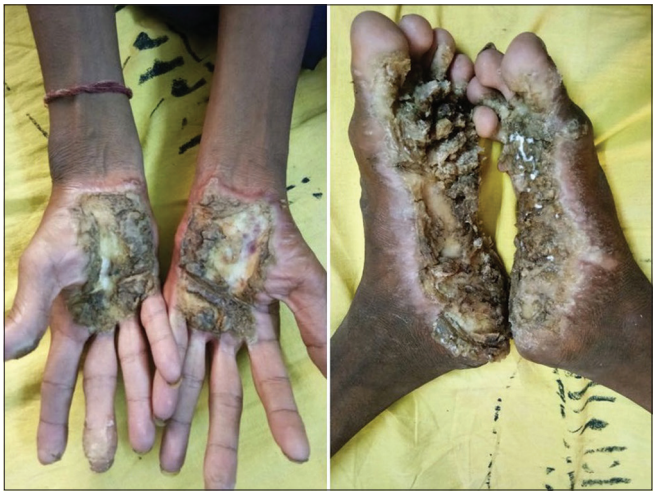

Figure 2: Palmoplantar keratoderma

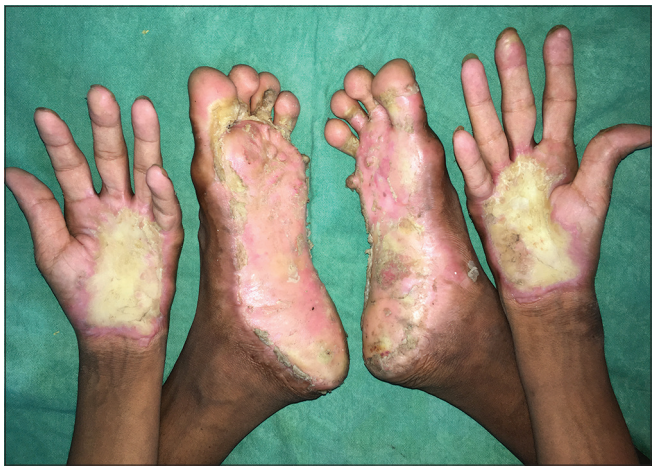

Figure 4: Partial improvement 2 months posttreatment

Nail changes such as dystrophic, lustreless, ridged, rough nails, abnormal curvatures, hyperkeratosis, onychogryphosis, onycholysis, subungual hyperkeratosis, and even absence of nails are often seen in OS patients.[2,6]

Short stature in our patient may be due to delayed physical development, as described earlier in OS patients.[2] Our patient seems to be of normal intelligence as that reported by Poulin *et al*.[7]

There is no satisfactory treatment of OS. Topical therapy with keratolytics, retinoids, wet dressing, and emollients give little relief.[1,7] Systemic retinoids has been reported to result in partial improvement but is ineffective in arresting the progressive course.[8] Our patient had temporary relief with capsule acitretin (0.5 mg/kg/day) along with topical keratolytics (20% urea cream) and emollients. For nonresponsive patients, surgical removal of hyperkeratotic plaque followed by skin grafting has been tried.[9]

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.

Conflicts of interest

There are no conflicts of interest.

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