INTRODUCTION

Olmsted syndrome (OS; mutilating palmoplantar keratoderma and periorificial involvement) is characterized by the presence of congenital, diffuse, symmetric, sharply margined, mutilating palmoplantar keratoderma with periorificial hyperkeratosis.\(^1\)

CASE REPORT

Three siblings of age 12, 10, and 6 years born of parents with consanguineous marriage, full term normal delivery with normal developmental milestones were brought with complaints of hair loss and palmoplantar keratoderma at the age of 2–3 years. No history of similar lesions in the family. Gradually lesions extended to knees and elbows.

On examination, children were moderately built and nourished. General and systemic examinations were within normal limits. There was palmoplantar keratoderma in all three siblings [Figures 1 and 2]. In two children, severe cheilitis, glossitis and keratotic plaques, and fissures around mouth were present. Scalp showed sparse hair with keratotic papule and follicular prominences, few scattered hair were thin and grew up to 1–2 cm in length [Figure 3]. Psoriasiform plaques were seen on elbows and knees. Paronychia and irregular dystrophy of finger and toe nails were present. The boy also had hypopigmented macules on the scalp. All routine blood and urine investigations were within normal limits.

Trichoscopy showed black dots, yellow dots in plenty. Tapered and thin hair, angulated hair, kinky gray hair, also had comma-shaped, “V” hair, broken hair, and vellus hair. The gap between hair follicles was more with follicular prominences [Figure 4].

DISCUSSION

OS is a rare congenital disorder characterized by palmoplantar and periorificial keratoderma, alopecia in most cases, and severe itching. Until recently, only 46 individuals had been reported, including 36 sporadic cases and four families.
Containing ten affected individuals. The definitive mode of inheritance was still uncertain, and autosomal-dominant, X-linked-dominant, and X-linked-recessive modes of inheritance had been proposed. Cambiaghi et al. reported transmission in two monozygotic male twins. They suggest that this condition is inherited as an X-linked dominant trait with reduced expression in female subjects. A case of a 6-year-old Indian girl in one report and two more unrelated female cases in another were reported. Whereas in our case, three siblings in a single family were affected, out of which two were females.

The diagnosis of the disease depends on clinical features. The two major ones are the symmetrical involvement of keratoderma of the palms and soles, and the symmetrical hyperkeratotic plaques around the body orifices. It starts in the neonatal period or in childhood. In our case, we have palmoplantar keratoderma, glossitis, cheilitis, and keratotic plaques around the mouth, sparse scalp hair with follicular prominences, hypopigmented macules over scalp and psoriasiform plaques were seen on elbows and knees. OS patients often show nail abnormalities including dystrophic, lusterless, ridged, rough nails, hyperkeratosis, onychogryphosis, leukonychia, irregular curvatures, onycholysis, paronychia, subungual hyperkeratosis, and even absence of nails. In our case, we had paronychia and irregular dystrophy of finger and toe nails.

Ogawa et al. report a case of this rare syndrome diagnosed in a 48-year-old woman, who developed several squamous cell carcinomas of limbs and adenocarcinoma of the lung.

CONCLUSION

We have reported this case to highlight the trichoscopy findings, female preponderance, and rarity in occurrence.

Financial support and sponsorship

Nil.
Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Dogra D, Ravindraprasad JS, Khanna N, Pandhi RK. Olmsted syndrome with hypotrichosis. Indian J Dermatol Venereol Leprol 1997;63:120‑2.
2. Elise Tonoli R, De Villa D, Hübner Frainer R, Pizzarro Meneghello L, Ricachnevsky N, de Quadros M. Olmsted syndrome. Case Rep Dermatol Med 2012;2012:927305.
3. Yaghoobi R, Omidian M, Sina N, Abtahian SA, Panahi-Bazaz MR. Olmsted syndrome in an Iranian family: Report of two new cases. Arch Iran Med 2007;10:246‑9.
4. Kumar P, Sharma PK, Kar HK. Olmsted syndrome. Indian J Dermatol 2008;53:93‑5.
5. Tharini GK, Hema N, Jayakumar S, Parveen B. Olmsted syndrome: Report of two cases. Indian J Dermatol 2011;56:591‑3.
6. Duchatelet S, Hovnanian A. Olmsted syndrome: Clinical, molecular and therapeutic aspects. Orphanet J Rare Dis 2015;10:33.
7. Ogawa F, Udono M, Murota H, Shimizu K, Takahashi H, Ishida-Yamamoto A, et al. Olmsted syndrome with squamous cell carcinoma of extremities and adenocarcinoma of the lung: Failure to detect loricrin gene mutation. Eur J Dermatol 2003;13:524‑8.