Net Letter

A family of Unna-Thost disease with one of them showing findings of epidermolytic keratoderma

Sir,

Palmoplantar keratodermas (PPKs) are a heterogenous group of disease. Inherited forms can occur with specific clinical and genetic findings (1). We report herein two cases with the diagnosis of Unna-Thost disease, one of them have atypical histopathological findings.

An 8-year-old male visited our clinic with thickening of palms and soles accompanied by marked hyperhidrosis and bad odor [Figure 1]. Family suffering of the disease is shown in the pedigree [Table 1]. On dermatological examination, yellowish hyperkeratosis and multiple pittings were found on the palms and soles. The oral mucosa, teeth and nails were normal. Histopathological examination of the biopsy taken from the patient's sole revealed orthokeratotic hyperkeratosis, hypergranulosis and acanthosis without epidermolysis. Interestingly, keratohyalin granules were increased in number, making it appear as if keratinization has started in the mid-epidermis. Also, some vacuolar degeneration in the granular layer was evident [Figure 2]. Hyphae were not detected on examination of the squames in the scraping of the soles with 10% potassium hydroxide.

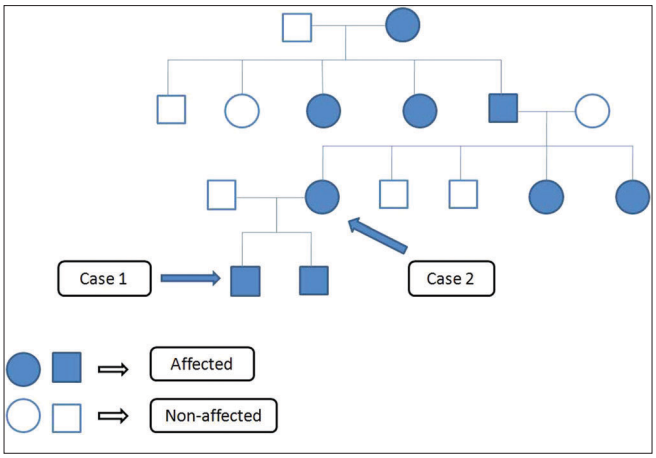

Table 1: Pedigree

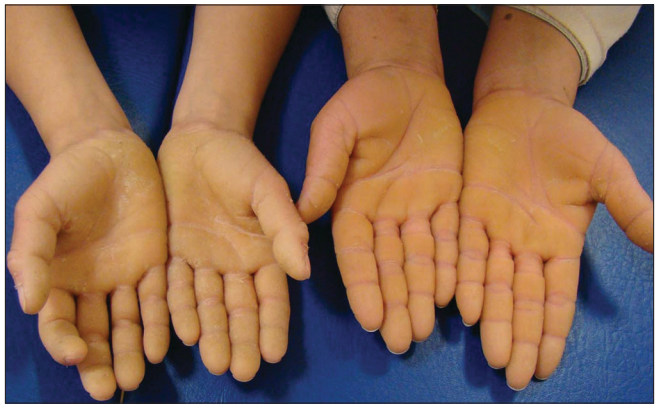

Figure 1: Palmar hyperkeratosis of case 1 and case 2

A 35-year-old patient, the mother of the patient in Case 1, had had thickening of the palms and soles since the age of 2 years. Hyperhidrosis was seen on the soles and palms. She had taken systemic acitretin and topical keratolytic drugs. On a dermatological examination, yellowish hyperkeratosis and desquamation were found on the palms and soles. Oral mucosa, teeth and nails were normal. Histopathological examination of the biopsy taken from the sole of the patient revealed orthokeratotic hyperkeratosis, hypergranulosis and acanthosis without epidermolysis [Figure 3a and b].

Hyphae were not detected on the soles. Topical tretinoin and moisturizing cream therapy were started

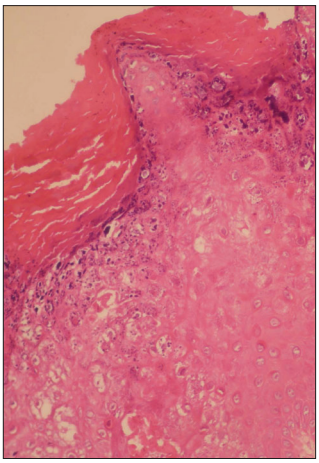

Figure 2: Histopathological section showing orthokeratotic hyperkeratosis, hypergranulosis, large keratohyalin granules and perinuclear vacuolization due to cellular degeneration (H and E, ×40)

How to cite this article: Karadag AS, Simsek GG. A family of Unna-Thost disease with one of them showing findings of epidermolytic keratoderma. Indian J Dermatol Venereol Leprol 2010;76:85.

Received: March, 2009. **Accepted:** August, 2009. **Source of Support:** Nil. **Conflict of Interest:** None declared.

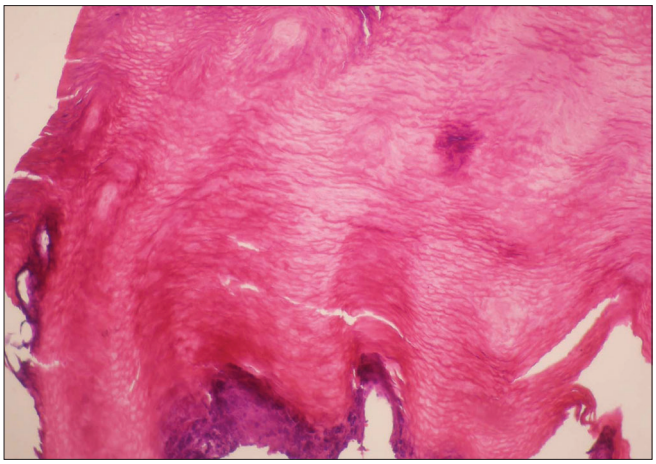

Figure 3a: There is massive orthokeratosis, hypergranulosis is seen in the mother's lesion (H and E, ×4)

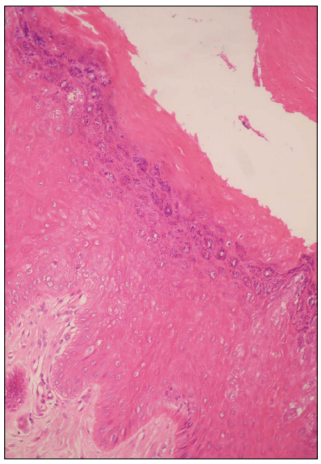

Figure 3b: Hypergranulosis, non-epidermolytic acanthosis area (H and E, ×40)

in both the patients and at present patients are under follow-up.

Unna-Thost disease (non-epidermolytic PPK) is inherited in an autosomally dominant manner, accompanied by diffuse PPK and without associated organ involvement. The disease developes in early childhood and persists throughout life. Clinically, there is hyperkeratosis on the palms and soles.[1]

Clinical features of epidermolytic PPK are very similiar to Unna-Thost disease. Differentiation from Unna-Thost disease can be made histopathologically, with the finding of epidermolysis.[2] Usual histological findings are orthokeratotic hyperkeratosis, hyper or hypogranulosis and acanthosis. These changes are non-specific and found in many types of keratoderma. There is no epidermolysis or vacuolar changes[1] in Unna-Thost disease.

In our cases, histopathological findings were consistent with the literature for Unna-Thost disease.[1] But, some vacuolar degeneration in the granular and dense globular keratotic areas in the spinous layer (also a finding of an epidermolytic PPK) were also seen in Case 1.[3]

Magro *et al*. reported that there was hypergranulosis in the Unna-Thost disease. However, the keratohyaline granules were distributed evenly in the cell and minute size and shape variations were found. They published that peripheral localization of the keratohyaline granules and perinuclear vacuolar change were not conspicious features. Also, in this article, large basophilic keratohyalin globules were seen in epidermolytic PPK. They recorded that a novel-type keratoderma that shared the features of both epidermolytic PPK and Unna-Thost disease. But, its genetic basis had not been determined.[4] Thinking of our cases, a finding of globular collections in Case 1 and not in Case 2 together with similiar clinical findings, we also suggest that all these findings support the diagnosis of Unna-Thost disease and that unusual findings can occur in the Unna-Thost disease.

Some authors think that epidermolytic PPK and Unna-Thost disease are identical to each other. Küster re-examined the Thost's family and he showed that they had histopathological findings of epidermolytic hyperkeratosis and both Thost's family and type Vörner had K9 keratin mutation.[4,5]

Histopathological findings are very important for the differential diagnosis of the epidermolytic PPK and Unna-Thost disease. But, there may be overlapping findings that result in diagnostic difficulties.

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DOI: 10.4103/0378-6323.58699 - **PMID**: 20061749

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