Corneal involvement in tyrosinemia type II initially mistaken for herpetic keratitis in an adult: a case report

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Background:

Tyrosinemia type II (Richner-Hanhart syndrome) is a rare metabolic disorder. Ocular involvement typically consists of pseudo-dendritic keratitis. It may be the presenting sign of tyrosinemia type II. Early diagnosis is essential because dietary modifications may result in a significant improvement of the signs of the disease.

Purpose : To report the case of a 40-year-old woman who presented with corneal lesions secondary to tyrosinemia type II, that were initially mistaken for herpetic keratitis.

Case Report :

A 40-year-old woman, with a history of mild mental retardation, presented with a painful red right eye. Marginal inferior corneal infiltration and ulcer were found. The patient received valaciclovir (2g a day), and then topical fluorometholone. Three months later, the patient consulted with bilateral ocular complaints. Slit-lamp examination revealed two corneal marginal ulcers in the right eye (RE) and a pseudodentritic inferior keratitis in the left eye (LE) (**figure 1**). Work-up showed a plantar hyperkeratosis (**figure 2**) and high serum tyrosine levels. The diagnosis of Richner Hanhart syndrome was made. The patient was prescribed tyrosine- and phenylalanine-restricted diet, and lubricants. Two months later, corneal lesions had healed (**figure 3**).

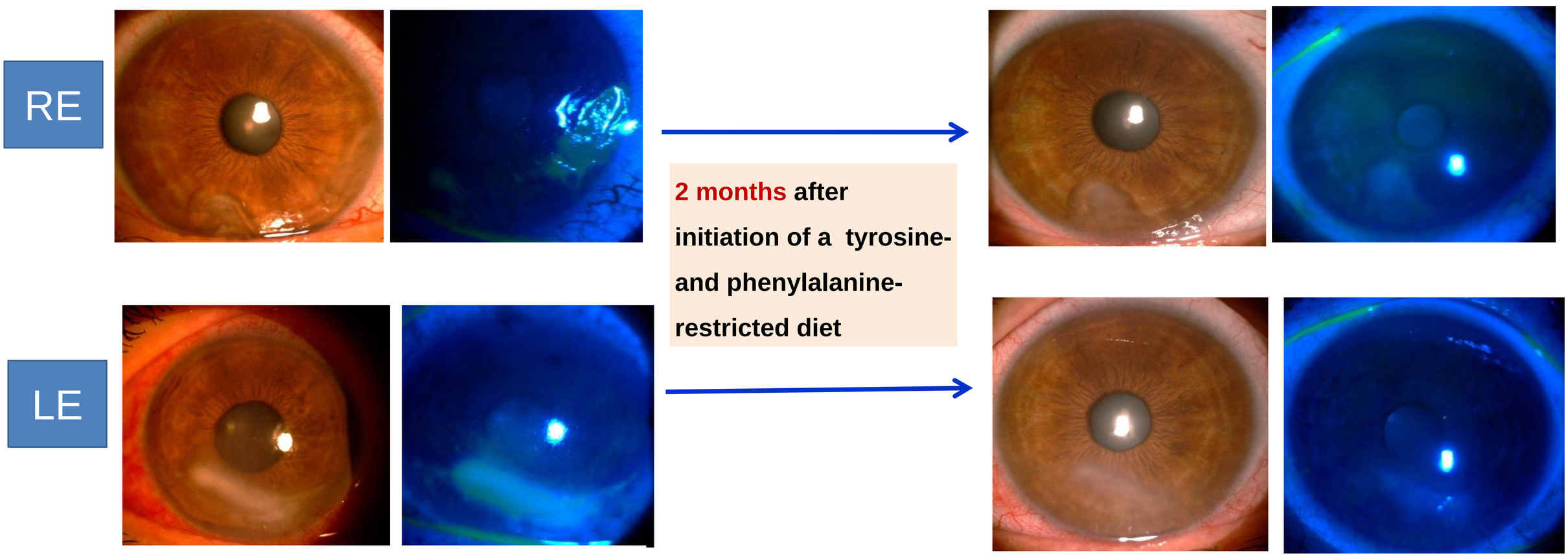


Figure 1: Slit-lamp photographs showing bilateral active pseudo-dentritic keratitis, staining with fluorescein of the RE and the LE

Figure 3: Healing of the corneal lesions

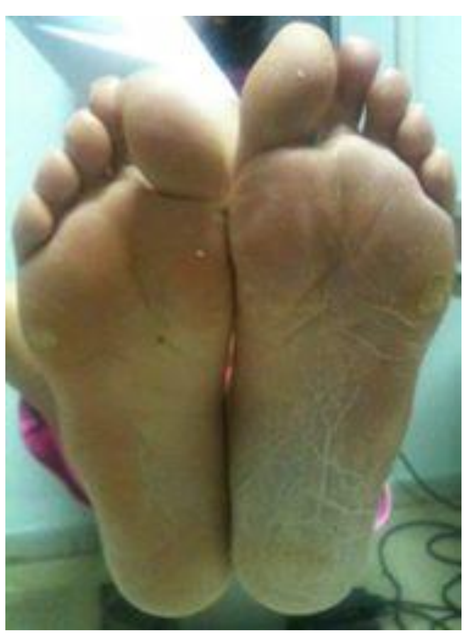
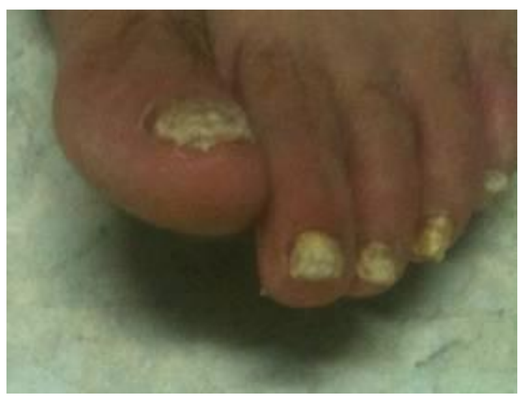


Figure 2: Plantar hyperkeratosis



Conclusions:

Richner-Hanhart syndrome should be suspected in patients with bilateral pseudodendritic corneal lesions unresponsive to antiviral therapy, even in adults. Systemic manifestations including palmoplantar keratosis and mental retardation should be looked for.