Images in Medicine

Neurotrophic keratitis in hereditary sensory autonomic neuropathy

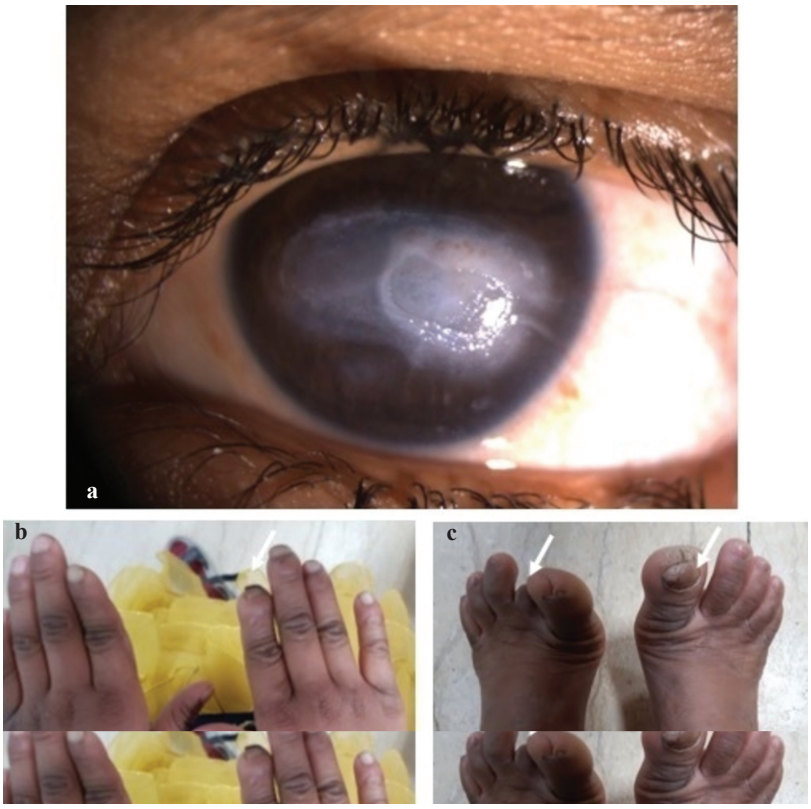

FIG 1. (a) Central 3 mm × 3.5 mm punched out corneal defect with surrounding 2 mm of infiltration; (b) multiple healed scarred lesions on all the finger tips; (c) toes with associated auto-amputation of two toes of left foot, with hyperkeratosis and fissure formation on tip of the right great toe

Hereditary sensory autonomic neuropathy-IV (HSAN-IV) is a rare autosomal recessive disorder that presents with congenital insensitivity to pain. This is usually associated with anhidrosis and dry eye. Poor pre-corneal tear film compromises the nourishment of corneal epithelium, predisposing it to recurrent erosions that heal poorly. A 6-year-old girl, known to be suffering from HSAN-IV presented to our outpatient department with complaints of diminution of vision of the left eye for 2 years. Visual acuity was 20/20 in the right eye and 2/200 in the left eye. Slit-lamp examination showed a central 3 mm×3.5 mm punched out defect with surrounding 2 mm of infiltration on the left eye (Fig. 1a). The other eye showed diffuse superficial punctate keratopathy. General physical examination revealed multiple healed scarred lesions on all the finger tips and toes with auto-amputation of two toes of left foot, with hyperkeratosis and fissure formation on tip of the right great toe (Fig. 1b and 1c). She was started on frequent topical lubricants and antibiotic drops. On 1-month follow-up, the corneal lesion healed with scarring. She was planned for left eye deep anterior lamellar keratoplasty.

Conflicts of interest. None declared

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