

A Rare Case of Thiel Behnke Corneal Dystrophy with Recurrence

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Abstract:

Thiel Behnke corneal dystrophy is an autosomal dominant corneal disorder¹. It is also known as honeycomb corneal dystrophy¹. A 28-year-old male came to OPD with chief complaint of defective vision in both eyes since childhood. On detailed ophthalmological examination, both eyes showed subepithelial irregular gray opacities arranged in honeycomb pattern, suggestive of Thiel Behnke corneal dystrophy.

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I. Introduction

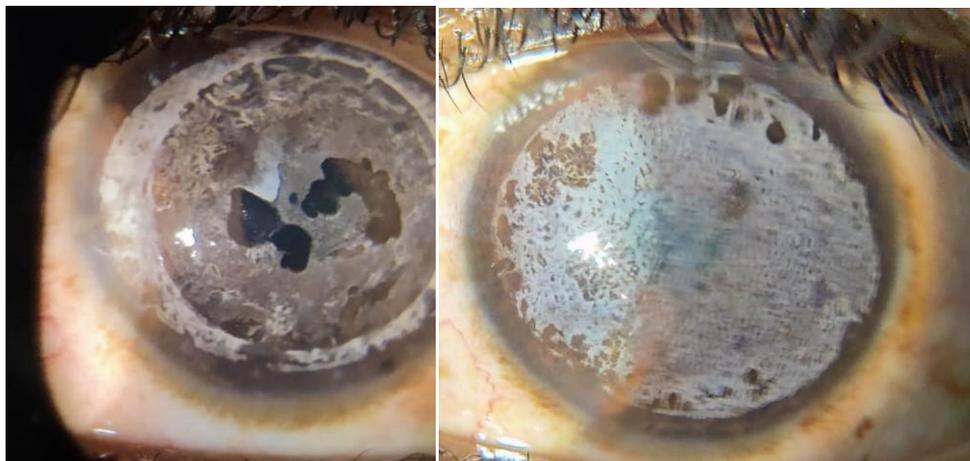
Thiel Behnke corneal dystrophy was first described in 1967^{1,2}. It is an autosomal dominant dystrophy which presents as honeycombed opacities and recurrent corneal erosions mainly affecting bowman's layer¹. It is difficult to differentiate clinically and histopathologically from reis buckler's corneal dystrophy^{3,4}. Initially corneal erosions are treated with topical antibiotics and artificial tears. Surgical treatment includes lamellar keratoplasty or phototherapeutic keratectomy or penetrating keratoplasty with chances of recurrence¹.

II. Case report

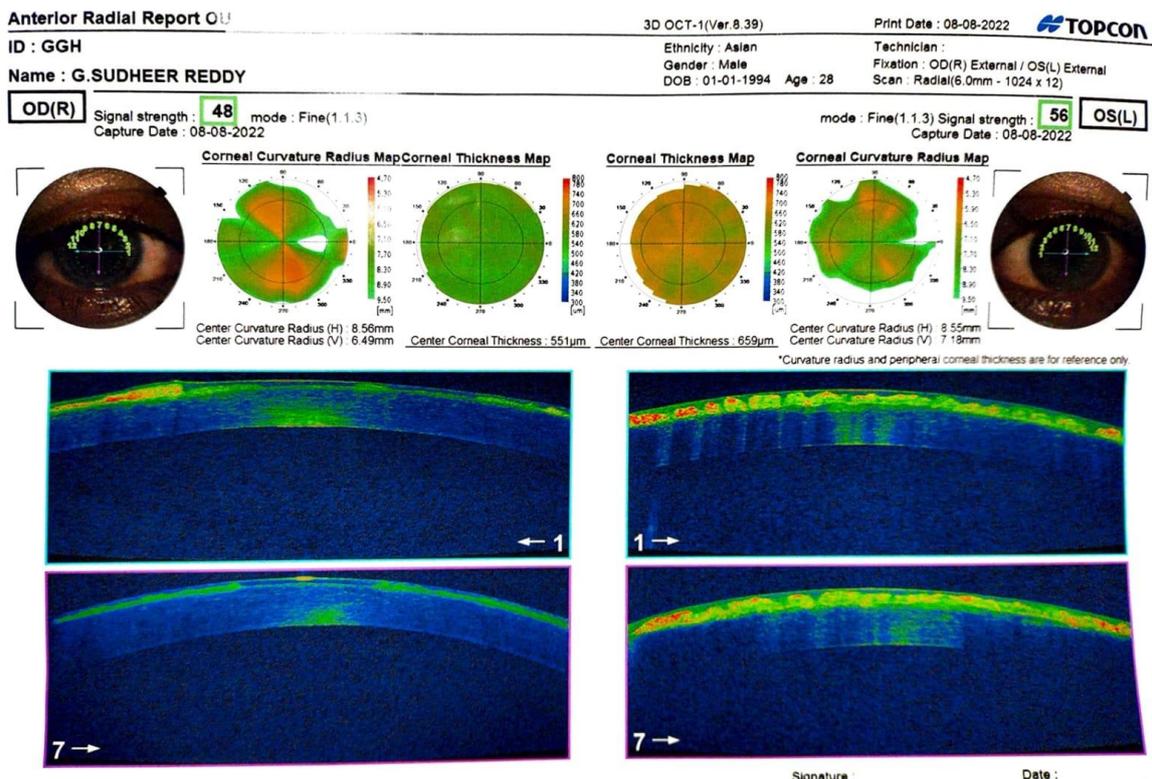
A 28-year-old male came to OPD with chief complaint of defective vision in both eyes since childhood. History of penetrating keratoplasty in RE and phototherapeutic keratectomy in LE 10 years back. Family history of presence of similar complaint in elder brother. His best corrected visual acuity in RE-6/24 and LE- counting fingers close to face.

Ocular examination:

Under slit lamp examination RE showed subepithelial irregular gray opacities arranged in honeycomb pattern predominantly at graft host junction. LE showed subepithelial irregular gray opacities in honeycomb pattern. Both eyes showed lucid interval between opacities and limbus. In both eyes clear stroma and endothelium is noted. Fluorescein stain is negative. Both eyes B-scan is normal.



AS-OCT both eyes showed thick band of saw tooth shaped pattern with moderate reflectivity at bowman's layer with clear stroma and endothelium.



A diagnosis of the Thiel Behnke corneal dystrophy was made.

III. Discussion

The bowman's layer is acellular, smooth, non-regenerating layer between epithelium and corneal stroma¹. Corneal dystrophies involving bowman's layer are¹

Type-1: Reis buckler corneal dystrophy (RBCD)

Type-2: Thiel Behnke corneal dystrophy (TBCD)

Reis buckler's corneal dystrophy is characterized by recurrent painful corneal erosions that often begin in first 1-2 years of life. It appears as gray white geographical dystrophy of bowman layer with normal stroma and endothelium.

Thiel Behnke dystrophy is a type of bowman's layer dystrophy also known as honeycomb dystrophy. It is a bilateral condition that has subepithelial opacities with autosomal dominant inheritance presenting in 2nd - 3rd decade of life with recurrent painful erosions and gradual decrease in vision.

Treatment initially includes topical artificial tears, patching, debridement or bandage soft contact lenses for corneal erosions. Topical hyperosmotic agents are better tolerated in recurrent corneal erosions. Surgical treatment includes lamellar keratoplasty or penetrating keratoplasty with chances of recurrence. Other modalities include Anteriorstromal micropuncture, excimer laser ablation (phototherapeutic keratectomy). Recurrences are common even after surgery.

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