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Second Re-emergence of Reis-Bücklers Corneal Dystrophy: Clinical and Histopathological Findings

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BACKGROUND

Reis-Bücklers corneal dystrophy (RB) is a rare, bilateral disease characterized by superficial gray opacities and progressive visual impairment. RB is caused by a specific mutation in the transforming growth factor beta induced gene and follows an autosomal dominant inheritance pattern with an incidence of <1% of all corneal dystrophies. This unique case discusses an RB patient with a history of lamellar keratectomy (LKP) OU 27 years prior, returning to the clinic for a second LKP due to re-emergence of the disease in the donor portion of the cornea. The purpose of this report was to assess this rare corneal disease clinically and analyze it histopathologically.

CLINICAL FINDINGS

A 41-year-old Hispanic female presented with a bilateral corneal dystrophy. Originally diagnosed at nine months of age, she reported experiencing several recurrent corneal erosions annually and progressively worsening vision OU. Best corrected visual acuity was 20/150 OD and 20/70 OS. Bilateral, asymmetric corneal opacities were noted at the level of anterior limiting lamina (ALL).

Biomicroscopy: Patchy, asymmetric, axial clouding of subepithelial tissue with accompanying epithelial abrasions were noted and consistent with the clinical manifestations of RB.

Histopathology: The diseased cornea revealed variable basal cell thickness and pallor. The basement membrane was irregular and lacked anchoring type VII collagen fibers. There was a total loss of ALL now replaced by a layer of fibrocellular connective tissue. Anterior stromal lamellae had signs of invading fibrocellular tissue at various depths and degenerating keratocytes throughout the cornea. These abnormalities were concentrated in the central cornea.

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CLINICAL IMAGES

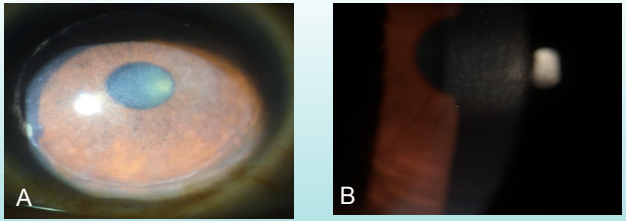

Fig 1. Anterior segment photo depicting the characteristic ground glass appearance of RB. (A) Gray, subepithelial opacities cloud the visual axis, reducing visual acuity. (B)

HISTOPATHOLOGICAL IMAGES

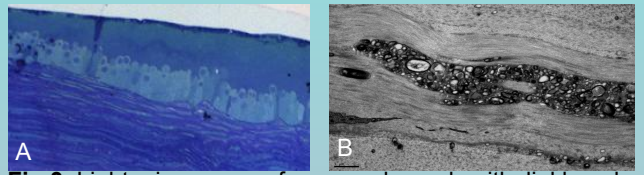

Fig 3. Light microscope of cornea showed epithelial basal cells with extreme pallor and and various thickness. The ALL was replaced by a layer of scar-like collagenous tissue. (A) (X 20) Electron microscope revealed invading electron dense fibrous tissue interdigitating into the anterior stroma. Numerous cells containing intracellular vacuoles were noted in the cross sections. (B) (X 9900)

DISCUSSION

This unique case discusses an RB patient with a history of LKP OU 27 years prior to returning to the clinic for a second LKP due to re-emergence of the disease in the donor portion of the cornea. The purpose of this report was to assess this rare corneal disease clinically and to analyze the consequences of the invasion of donor tissue histopathologically. Contradicting earlier literature, our histopathological assessment revealed that the ALL and stroma are both disrupted by fibrous tissue and active phagocytic cells. The epithelial fragility in RB is due to the loss of its foundation, ALL, explaining the frequent abrasions, erosions, and ocular discomfort.

Reis-Bücklers corneal dystrophy is known to re-emerge after transplantation because the surgery does not completely eliminate the underlying pathology. It should be emphasized to patients with RB or other corneal dystrophies that ultimately the disease will enter the healthy donor cornea and possibly necessitate an additional future keratectomy. Indeed, our patient is facing the possibility of a third transplant.

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