Is the lattice dystrophy of the cornea due to developmental anomalies of neural crest cells during embryogenesis (neurocristopathies)?

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SUMMARY

With the present paper authors try to give an embryological approach in understanding etiopathogenesis of lattice corneal dystrophy (LCD). The case of a 41-years-old man affected by isolated LCD has been examined at the Ophtalmologic Clinic of the University of Catania, Italy. Sore cornea has been excised during cornea transplantation, and it has been examined by transmission electron microscopy (TEM). Results confirm classic submicroscopic findings described in literature. However these findings offer possibility to consider an embryological interpretation of LCD pathogenesis. Our findings with those of literature put forward hypothesis of a hereditary etiopathogenetical role exerted by keratocytes, direct derivatives of neural crest cells during embryogenesis, probably via an abnormal gene expression in producing proteinaceous precursor of amyloid substance (APP). LCD could be considered a neurocristopathy.

INTRODUCTION

Since first reports of Biber (1899), Haab (1899) and Dimmer (1899), lattice cornea dystrophy (LCD) has been identified as a degenerative corneal disease supported by a dominant autosomal inheritance. In agreement with most of literature reports, in clinical history of our own case, object of present report, first clinical signs of disease, corneal erosions and progressive limitations of vision, begin at the end of the first decade of age, and they are based on presence of small, grey ribbons and spots located in the anterior layers of cornea. Up to date there are a lot of histological observations, ultrastructural findings, and immunocytochemical stud-