Pathology of Hereditary Juvenile Retinoschisis

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The clinicopathologic data are presented in one familial case and in two other cases of juvenile retinoschisis in which the family history was not available.

The principal feature in all three cases is a large retinal schisis cavity, the anterior wall of which has become a retrolental membrane that is responsible for the clinical picture of leukokoria. The schisis is seen histo-logically as a splitting within the nerve fiber layer. It appears impossible to state whether the changes found in the pigment epithelium of the posterior pole are secondary to the changes in the sensory retina.

Discussion

Wynkelman: What was the reason for the enucleation in the American cases you have mentioned? I do not believe that this disease leads to a condition that necessitates enucleation.

Manschot: The leukokoria symptom. Juvenile retinoschisis belongs to the group of diseases that are summarized under the heading of pseudoretinoblastoma.

Hamburg: As I understand it, the schisis may penetrate into various portions of the internal layers. What is the concept of the internal limiting membrane? As far as I am aware, there is as yet no agreement concerning the question of what forms this membrane: the retina? The external layer of the vitreous humor? Or these two together?

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Manschot: According to Fine, the internal limiting membrane is a basal membrane formed by the Müller cells. Thin collagenous fibers of the vitreous humor are tightly attached to it.

Deutman (Rotterdam): You suggest in your first slide that juvenile retinoschisis always has a sex-linked heredity. As a rule

the heredity of juvenile retinoschisis is indeed sex-linked but occasionally

patients present an autosomally recessive juvenile retinoschisis [Cibis

1965; personal observations]. In these cases there are no foveal alterations as a rule. Is the histology of this condition also known?

Manschot: Thank you for this supplementary information. I am not familiar with the histology of this condition.