Tapetum lucidum in a Carrier of X-Chromosomal Atrophia retinae pigmentosa

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Demonstration, in a family with X-chromosomal atrophia retinae pigmentosa of a carrier who exhibited the typical fundus image of a tapetum lucidum reflection: a brilliant radiance of the retina during mirror examination, reminiscent of the tapetum lucidum in certain nocturnal animals. This so-called tapetal reflex in humans, described for the first time by Ida Mann in 1937, is supposed to be more or less pathog-nomonic of carriers of the X-chromosomal form of retinitis pigmentosa. The tapetum lucidum is observed as immobile, gold-colored reflections, which, when examined with the fundus contact glass, exhibit a somewhat coarse granular appearance and are localized deep in relation to the related vessels. The morbid-anatomical substrate is unknown.

Ricci et al. in 1963 called the attention to the fact that in their cases the reflection disappeared after protracted illumination, and reappeared after a period of dark-adaptation. A related phenomenon can be observed in Oguchi's disease, in which the abnormal, greyish-white color of the retina disappears after a stay in the dark: the so-called phenomenon of Mizuo. In tapetum lucidum the inverse occurs; accordingly, it is sometimes called an 'inverse Mizuo phenomenon'. Our patient, a girl whom we saw for the first time in 1963 when she was 10 years old, exhibited a distinct tapetum lucidum reflection over the entire retina, most pronounced in the foveal area. The visual acuity was subnormal: right eye, 6/10 after correction and left eye, 8/10 after correction. Subjectively there was marked night blindness, although objectively no abnormalities of the dark-adaptation curve could be demonstrated; the visual fields were intact, and the ERG was slightly abnormal. There was a red-green disorder of the color sense of the acquired type.

248

Breebaart-de Miranda

During a follow-up period of 10 years, with an annual examination, the following additional observations were made. Firstly, we have not observed the inverse Mizuo effect. Secondly, with the passage of years -the patient is 20 years old at the time of writing - the tapetum lucidum reflection gradually grew less pronounced and currently is hardly visible in the right eye. It is interesting to note that parallel to this fading, the functional disorders gradually improved: the visual acuity increased, with practically unchanged refraction, to OD 8/10 and OS 12/10. The subjective symptom of night blindness has also disappeared entirely, and the ERG is currently normal. The EOG presents no abnormalities, only the disorder of the color sense remained unaltered.

No satisfactory explanation of the disappearance of this tapetum lucidum, associated with functional recovery, can be given. The precise anatomical localization of the anomaly cannot be

determined with certainty. It seems plausible that the anomaly is situated in the choroid or Bruch's membrane, although a primary localization in the pigmented epithelium cannot be excluded, in view of the results of recent studies of the pathophysiology of the pigmented epithelium and its role in the various forms of tapeto-retinal degeneration.