Gyrate Atrophy and Choroidal Neovascularization

A 53-year-old, high-myopic white man affected with gyrate atrophy (GA) was referred in May 1983 because of a sudden loss of vision in his left eye. His best-corrected visual acuities were 20/60 OD and 20/400, with a central scotoma, OS. On ophthalmic examination, peripheral and peripapillary chorioretinal atrophy was seen in both fundi. A localized neurosensory detachment with choroidal neovascularization (CNV) was suspected in the left eye. Conventional fluorescein angiography showed a well-defined subfoveal CNV (Figure 1). Because of the central localization of CNV, no laser treatment was advised.

In May 1995, the patient's visual acuity was limited to hand movements OU. Ophthalmoscopy of the left eye showed chorioretinal atrophy involving the central macula. Below a small, subfoveal, and apparently fibrotic scar, a triangular granular pigmented area was seen. Fluorescein angiography still revealed leakage from the central subretinal lesion, surrounded by atrophy of the retinal pigment epithelium and choriocapillaris (Figure 2).

COMMENT

In their study of 29 patients affected with GA, Takki and Milton concluded that the decrease in visual acuity was dependent primarily on atrophic macular involvement and cataract formation. To our knowledge, no previous cases of GA associated with CNV have been reported to date.

High myopia has to be considered as a typical feature of patients affected with GA. In a recent study of the clinicopathologic features of 123 surgically removed subretinal membranes, Grossniklaus et al concluded that CNV represents a stereotypical, nonspecific response, regardless of the underlying disease. If one accepts those conclusions, the same pathogenetic course of CNV as in pathologic myopia might be postulated in our patients affected with GA. In both conditions, large areas of chorioretinal atrophy with a relative ischemia-hypoxia condition are present.

Excluding any further complication, this rare, inherited, chorioretinal dystrophy has an unfavorable prognosis. Because CNV might occur in patients affected with GA, further loss of vision may be experienced during their lifetime.

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REFERENCES


Figure 1. Conventional early-phase (left) and late-phase (right) fluorescein angiography performed in May 1983 shows a well-defined choroidal neovascularization in the patient's left eye, contiguous to the temporal border of the peripapillary chorioretinal atrophic area.

Figure 2. Conventional fluorescein angiography performed 12 years later shows some still-evident subretinal leakage from the partial fibrotic scar.