LATE RADIATION RETINOPATHY IN A PATIENT TREATED FOR RETINOBLASTOMA

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Summary. We report a case of a young man who developed radiation retinopathy 12 years after irradiation for retinoblastoma. With a history of bilateral retinoblastoma, our patient presented clustered retinal teleangiectasias in the non-enucleated but treated with radiation eye. The lesions were associated with hemorrhages and gradually increasing retinal exudation that deteriorated best corrected visual acuity (BCVA) from 0.25 to 0.1. Detailed ophthalmic examination was performed incl. fluorescein angiography (FA), indocyanine green angiography (ICG) and optical coherence tomography (OCT). The ICG highlighted the vascular anomalies that were clearly hyperfluorescent in the late phase. After 3 sessions of focal ALC the outlined edema on Oct scans diminished and BCVA improved to 0.2. Some retinoschisis-like cystic changes remained on OCT retinal scans. The manifested changes had common features with those found in diabetic retinopathy. ICG staining of the retinal vascular anomalies was found. Radiation retinopathy may occur more than a decade following radiation treatment for retinoblastoma. Appropriate laser treatment may overcome the advancing process.

Key words: *radiation retinopathy, OCT, ICG, retinoblastoma*

INTRODUCTION

Radiation retinopathy is a slowly progressive disease with a delayed onset. The alterations in the structure and permeability of retinal blood vessels usually occur months to years after exposure to radiation [3]. It was reported to develop in 12% [1] and 24% [4] and was first detected 11-72 months (mean 37 months) after irradiation therapy [1] for retinoblastoma. However we present a patient whose initial clinical signs appeared 12 years (144 months) after ionizing radiation treatment.

CASE REPORT

A 16-year-old patient, previously irradiated for retinoblastoma, was referred for vascular anomalies in the right eye. The latter were observed for about 3 years but appeared to increase. At the age of 9 months, a bilateral retinoblastoma had been diagnosed. The left eye with a large tumoral mass was enucleated and the pathologic examination confirmed the diagnosis. The right eye presented two foci and was irradiated with 3650 cGy (3x/week 365 cGy; 8MeV). A supportive chemotherapy was additionally given. Both lesions subsequently regressed, the small one as a sharply demarcated zone of chorioretinal atrophy temporal of the fovea and the larger one as an atrophic scar inferior to the optic disc with two foci of calcification.

His best corrected visual acuity was 20/80, the anterior segment was unremarkable. The retinal pigment epithelium was minimally pigmented and the choroidal vascular pattern could easily be seen. Two atrophic chorioretinal scars, corresponding to the tumoral lesions, were present. Two clusters of retinal micro-aneurysms and/or dot retinal hemorrhages were found, one temporally and another superotemporally of the macular scar as well some scattered similar lesions. No retinal exudates or larger retinal hemorrhages were seen. Fluorescein angiography (FA) revealed filling of some of these lesions, in others fluorescence was blocked by associated intraretinal hemorrhages or the lesions were thrombosed (Fig 1A). Retinal ischemia was not observed, neither in the macular area, nor in the periphery.

Two years later, his visual acuity did not changed, however larger retinal hemorrhages appeared as well as retinal exudation superior to the macular atrophic scar (Fig.1B). A normal pattern of infrared autofluorescence was seen apart from masking by the calcifications in the regressed tumor scar. Indocyanine green (ICG) angiography highlighted better the vascular anomalies than FA, which became hyperfluorescent in the early venous phase (Fig. 2A). In the late phase, their walls remained hyperfluorescent (Fig. 2B). The atrophic scars were hypofluorescent while the retinal exudates blocked choroidal fluorescence. The hemorrhages only caused a minor masking phenomenon. An ill-defined area of hyperfluorescence was seen superotemporally to the disc in the late phase.

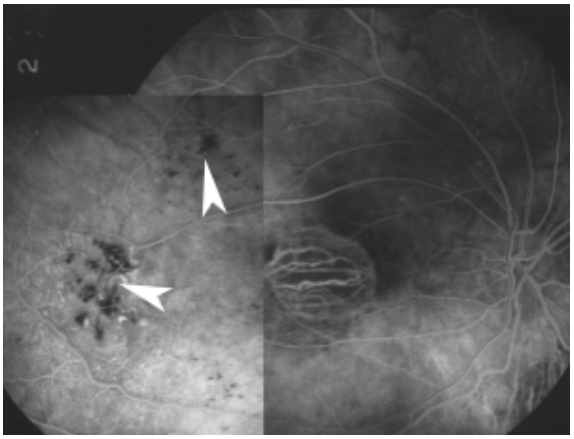

Fig. 1. FA (venous phase) at the age of 16 years showing two clusters of retinal vascular abnormalities (white arrowheads) and a perifoveal area of well-demarcated chorioretinal atrophy at the place of the regressed tumoral lesion

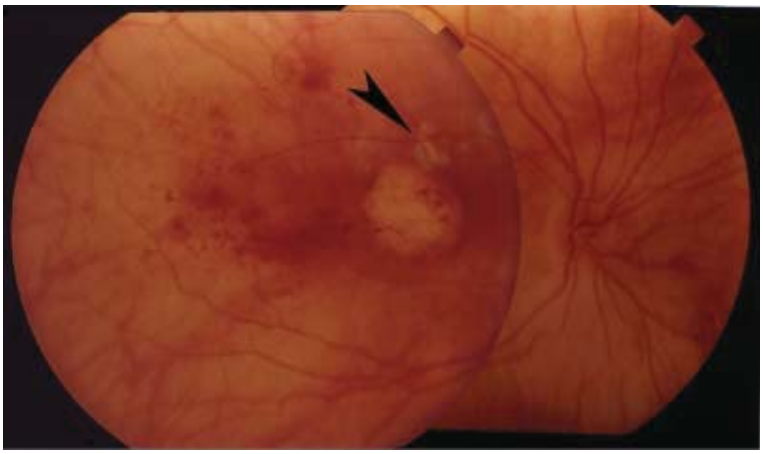

Fig. 2. Fundus photo at the age of 18. Note the occurred retinal exudation above the macular scar (black arrowhead). The vascular abnormalities have increased

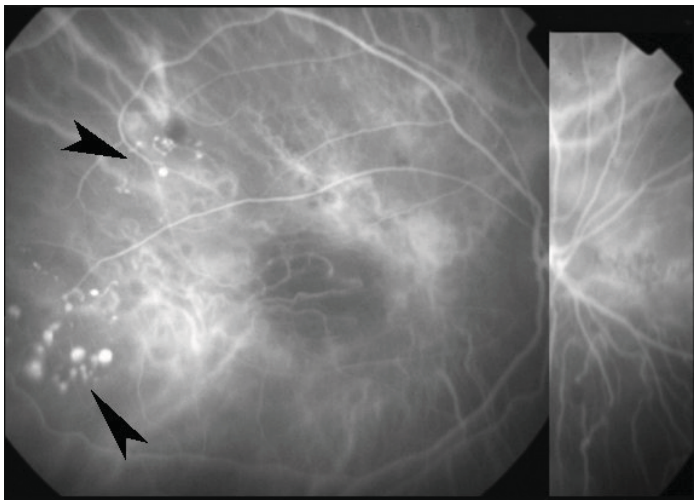

Fig. 3A. Early venous phase ICG angiogram at the age of 18 years showing hyperfluorescence of the vascular abnormalities (black arrowheads)

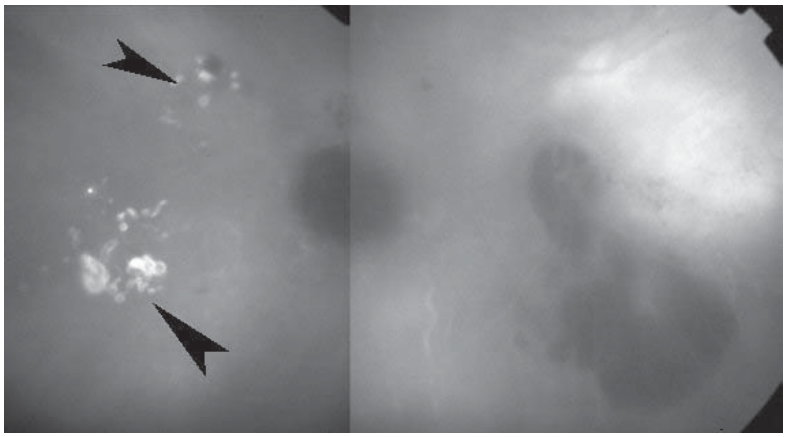

Fig. 3B. Late phase ICG angiogram of the posterior pole – remark two obvious clusters of vascular lesions (black arrowheads), the hypoflurescent chorioretinal scars and masking of exudates above the fovea

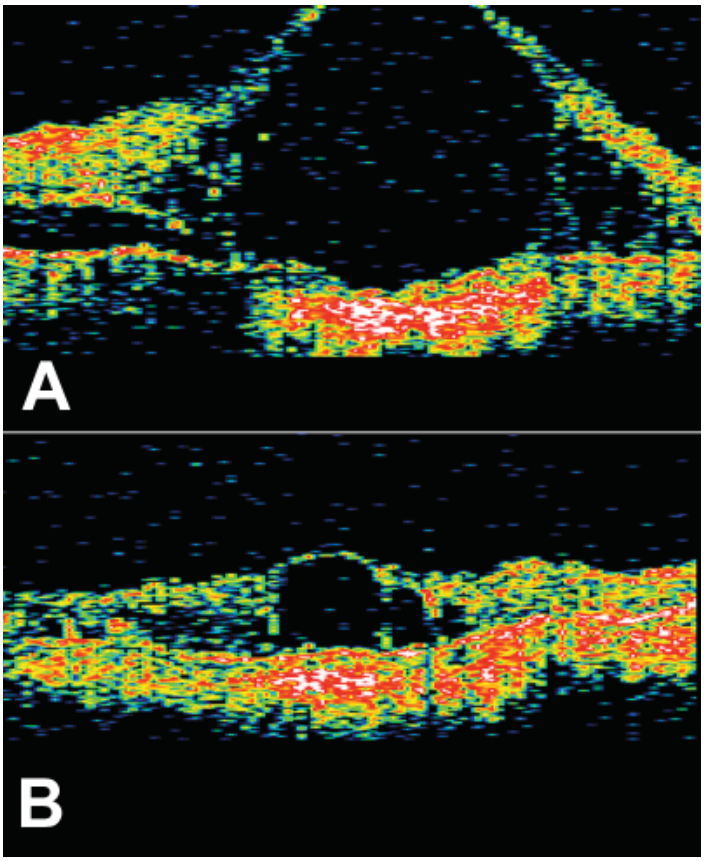

Fig. 4. A – OCT scan through the macula at the age of 19 years with severe macular edema; B – OCT scan 9 months later, after 3 sessions of focal argon laser treatment. Note the substantially decreased edema and some residual cystic changes

Almost a year later, his visual acuity decreased from 20/80 to 20/200. Optical coherence tomography (OCT) revealed severe retinal edema in the macular area (Fig 3). After three sessions of focal argon laser photocoagulation his best corrected visual acuity improved to 20/100 and the macular edema on OCT regressed. On follow-up OCT sessions persisting intraretinal cystic changes were recorded in the macula.

DISCUSSION

Alterations of retinal vessels, such as microaneurysms, dilated capillaries, cotton-wool spots, perivascular sheathing, irregular loss of retinal capillaries, hemorrhages and hard exudates become apparent months or several years after X-ray irradiation of the orbital region. The vascular changes are similar to those found in diabetic retinopathy. Radiation retinopathy threshold depends on the delivered total dose, irradiated retinal area and fractionation scheme. Retinal damage usually develops after 3000 to 3500 cGy and may be enhanced by simultaneous chemotherapy.

This patient developed clustered retinal telangiectasias with intraretinal hemorrhages 12 years after irradiation for retinoblastoma at the age of 9 months. The number of dilated capillaries gradually increased. His visual acuity deteriorated because of intraretinal accumulation of fluid, documented by OCT and formation of hard exudates. We consider these lesions to be a late radiation effect although FA failed to demonstrate major capillary dropout – the hallmark of radiation-induced retinopathy [2]. The retinal vascular anomalies were strikingly highlighted by ICG angiography: they appeared as highly fluorescent dots in the early venous phase, while in the late phase their wall was stained. The regression of the macular edema after appropriate focal laser treatment, as confirmed by OCT scans, was impressive.

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