Retinal Diseases
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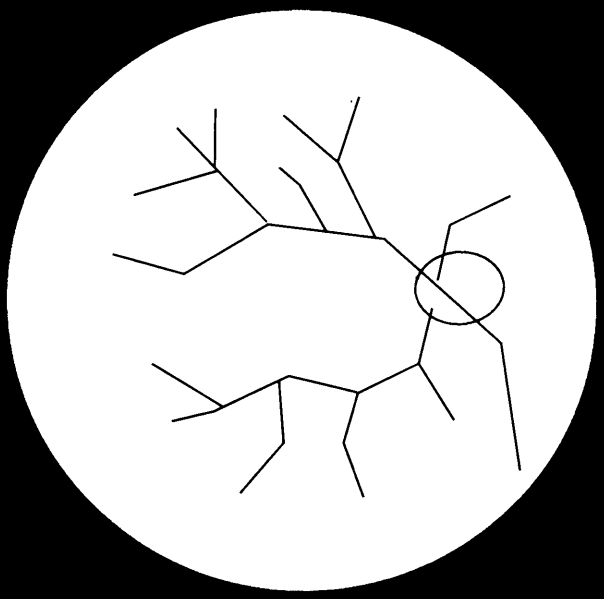

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REPRINTS

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OPTIC DISC CAPILLARY HEMANGIOMA
Fluoroangiographic and echographic study of two cases

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Abstract

Optic disc capillary hemangioma together with racemose aneurysm and cavernous hemangioma is a rare hamartoma of the optic nerve head. In two 30- and 72-year-old patients fluorescein angiograms showed highly vascularized lesions partially covering and surrounding the optic disc with early fluorescein injection and late marked leakage. Standardized A-scan echography displayed spontaneous vertical movements of the high reflective echoes from the solid lesions and normal retrobulbar optic nerve widths. On the basis of echographic results choroidal melanoma involving the optic nerve was excluded. B-scan examination and computerized tomography nicely evidenced the lesion in one case. A clinical diagnosis of unilateral disc capillary hemangioma was made in both patients. Xenon photocoagulation of the hemangioma was performed in one eye with exudative macular detachment. No tumor growth was observed after about two year follow-up in both patients.

Introduction

According to Gass and Braunstein,[1] capillary hemangiomas are usually easy to identify when they protrude from the anterior surface of the optic disc and juxtapapillary retina (endophytic tumors) but are usually misdiagnosed as papilledema, papillitis, choroiditis, choroidal neovascularization or choroidal hemangioma when they develop within the middle and anterior layers of the juxtapapillary retina (sessile or exophytic tumors). We report the clinical features of two patients with endophytic and sessile capillary hemangioma.

Case reports

Case 1. A healthy 30-year-old patient who complained of progressive unilateral visual loss was examined at the University Eye Clinic of Padua. On initial examination visual acuity was 20/100 in the right eye and 20/20 in the left eye. The adnexa, anterior segment and IOP were normal in both eyes.

Funduscopy revealed a red sponge-like roundish solid lesion sized about two disc diameters and elevated five diopters over the nasal half of the optic disc in the right eye.

Serous detachment of the retina surrounding the optic disc and several hard exudates were located both in the macular area and nasally to the optic disc. Fluorescein angiography revealed early lobular filling of the tumor in the arterial phases; dye leakage from the tumor and pooling of the surrounding subretinal space were observed in the late phases (Fig. 1).

Fluorescein angiography of the left eye was normal. Contact B-scan echographic examination clearly showed a small lesion overlying the optic disc (Fig. 2); standardized A-scan examination revealed high reflective echoes (with spontaneous vertical movements at T-20 dB) from the lesion elevated 2.5 μsec (Fig. 3) and a normal retrobulbar optic nerve thickness. A disc-related superior arcuate field defect was plotted by Goldmann perimetry in the right eye. The small tumor protruding from the optic disc could also be detected with cerebral computed tomography; no extraocular extension or brain lesions were detected on contrast enhancement.

General physical and neurological examinations and routine laboratory analyses were negative. Direct treatment of the angioma with moderately intense applications of xenon photocoagulation resulted in

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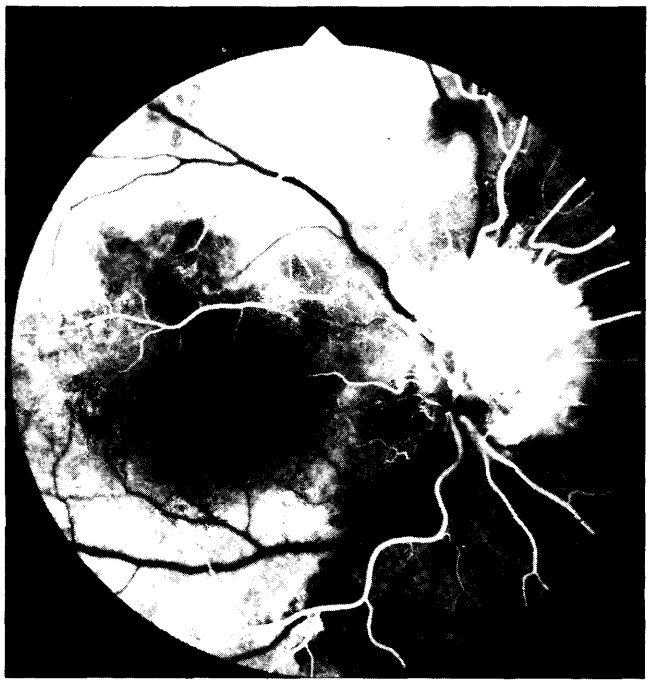

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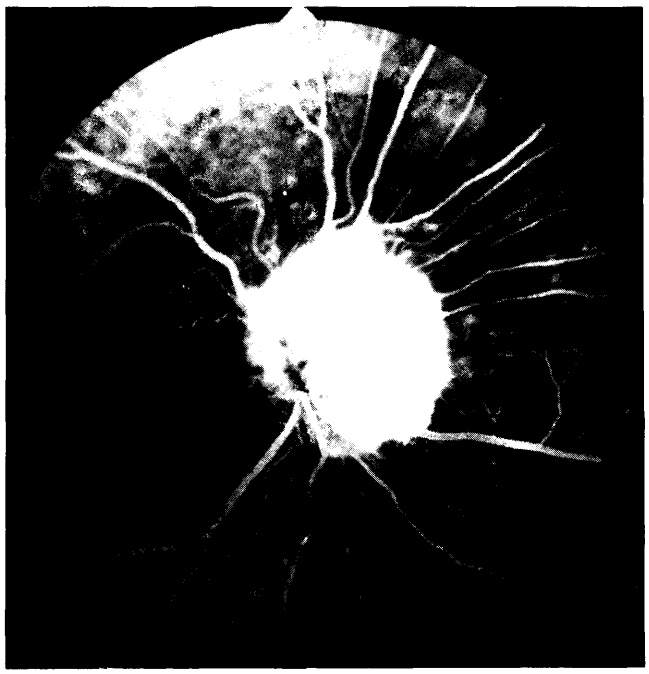


B

Fig. 1. Case. 1. Fluorescein angiography: during the arterial phase a lobular filling of the tumor is evident (A). Later on the dye markedly stains the mass and the surrounding retina (B).

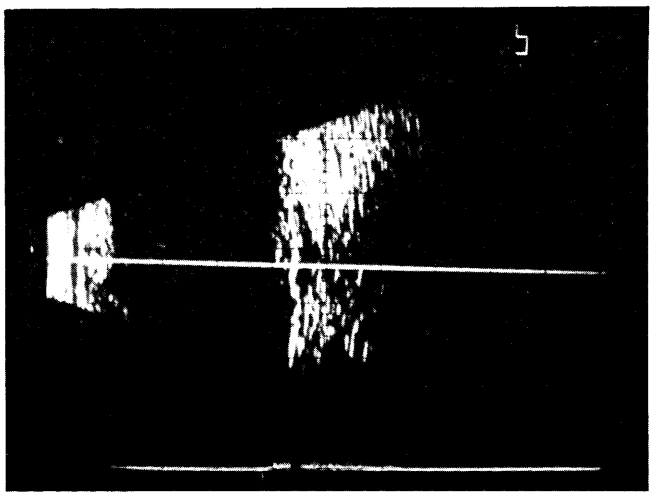


Fig. 2. Case 1. B-scan echogram showing the small mass overlying the optic disc.

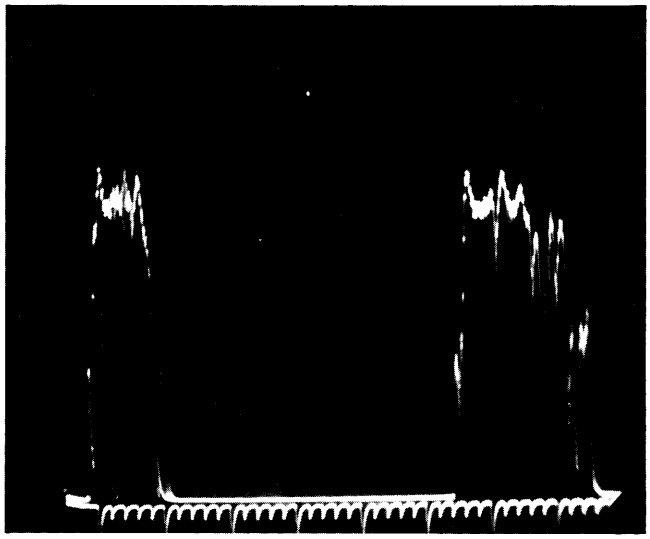
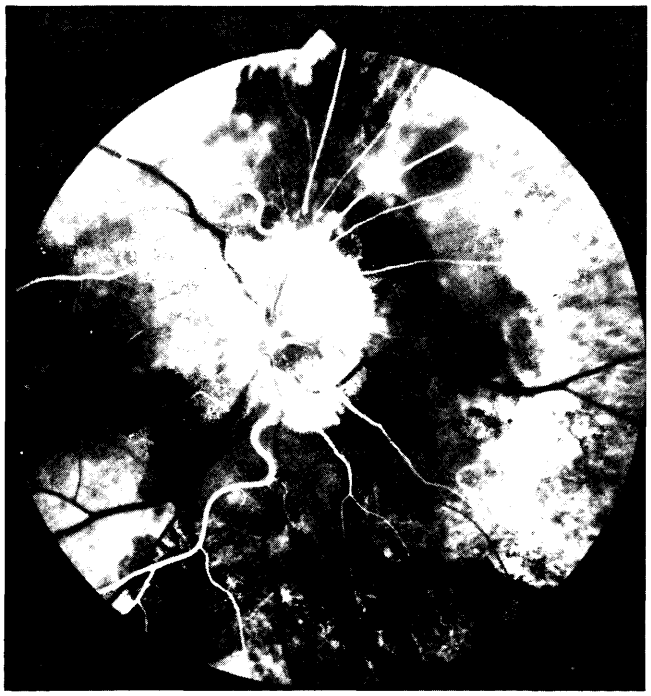


Fig. 3. Case 1. Standardized A-scan echography reveals high reflective echoes from the lesion without acoustic shadowing.

small vitreous hemorrhage which cleared after about two months. During the two-year follow up visual acuity decreased to 20/200.

The area of subretinal exudation including the macula enlarged and was surrounded by yellow circinate exudate inferiorly; radial folds of the internal limiting membrane in the papillo-macular bundle area, and gliosis beneath the inferior margin of the optic disc were evident. The tumor flattened slightly but little or no change was observed in follow-up fluorescein angiograms except a reduction of the hyperfluorescent area in late phases (Fig. 4). No further photocoagulations were performed.



A

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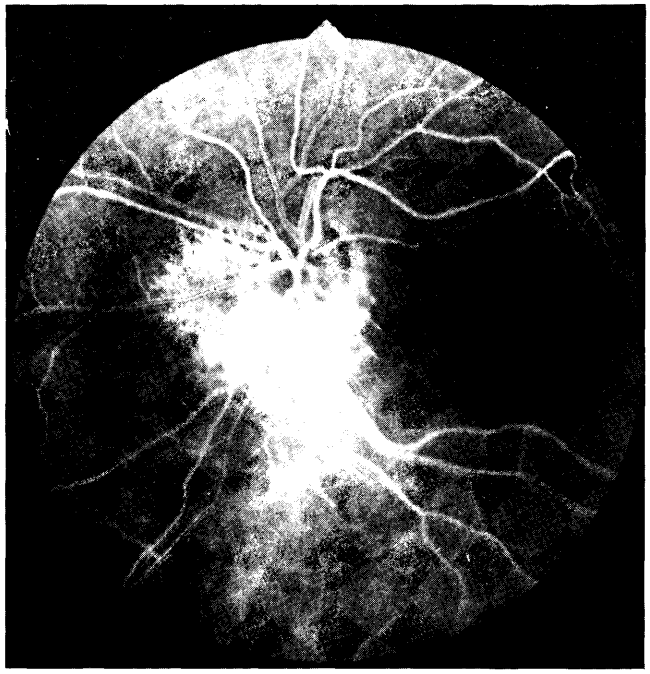
Fig. 4. Case 1. Fluorescein angiography after treatment: in the arterial phase a slight reduction in the extension of the tumor can be observed. Mild stretching of the surrounding retinal vessels due to gliosis is also evident (A). In late phases patchy leakage still appears (B).



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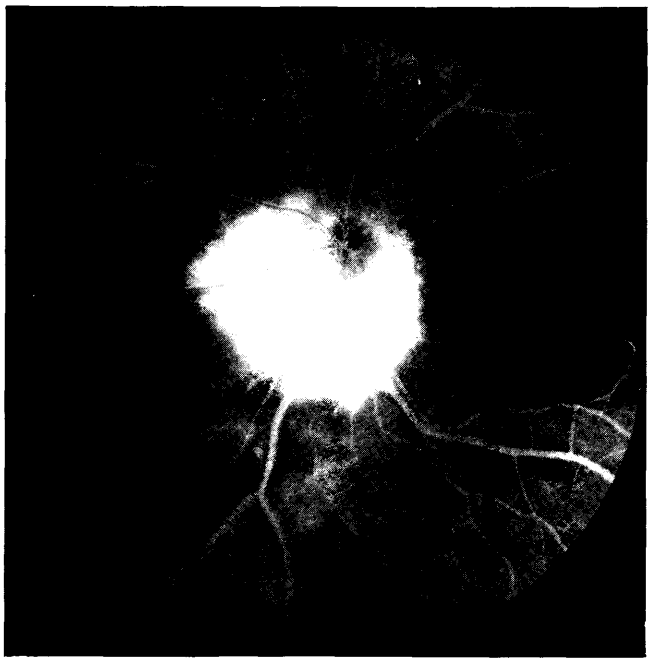

B

Fig. 5. Case 2. Fluorescein angiography: in early phases lacy hyperfluorescence appears infero-nasally to the disc (A). Deep leakage of the dye involves the tumor and the surrounding retina (B).

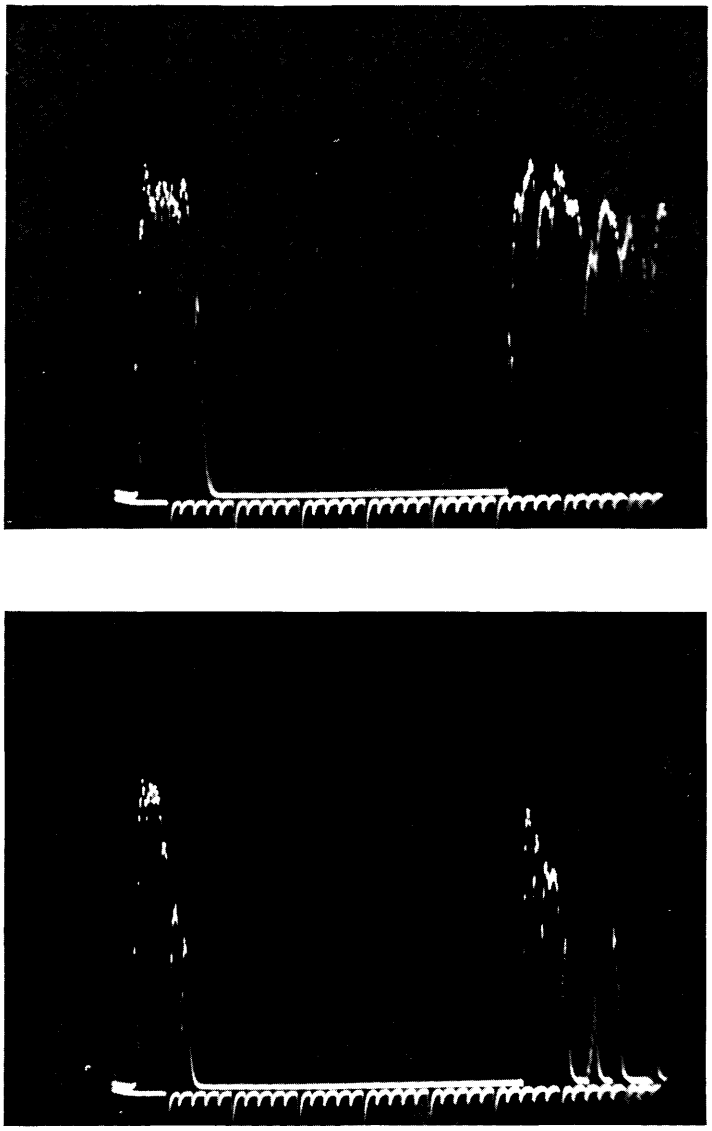

Fig. 6. Case 2. Standardized A-scan echography shows a high reflective lesion without acoustic shadowing at the posterior pole; (A) spontaneous vertical movements of subretinal echo spikes could be observed at reduced system sensitivity (B).

Case 2. A sudden decrease of vision and a tentative diagnosis of impending retinal vein occlusion was reported in the left eye of a 72-year-old woman, two years prior to admission to the University Eye Clinic of Padua. Her medical history revealed surgery for thyroid goiter. The patient had been suffering from angina pectoris for four years. Results of general physical and neurologic examinations were normal. Corrected visual acuity was 20/20 in the right eye and 20/400 in the left eye. As in Case 1 adnexa, anterior segment and IOP were normal in both eyes. Visual field examination was normal in the right eye and showed central hemianopic temporal scotoma and enlarged blind spot in the left eye. Biomicroscopic and ophthalmoscopic examination of the left eye revealed blurring of the nasal and inferior margins of

the optic disc, and elevation (4D), extending over half a disc diameter, of the retina around the nasal and inferior side of the optic nerve head.

Mild dilatation of the superficial capillaries resembling striate hemorrhages was observed in the raised retina but there were no exudates at the posterior pole. Fluorescein angiograms revealed a sessile juxtapapillary capillary angioma which perfused during the retinal arterial phase (Fig. 5).

During the late phases extensive leakage of dye from the tumor vessels was observed. The small juxtapapillary lesion was difficult to detect with B-scan echography; a solid (no after movements) retinal detachment elevated 2 μsec with high reflective internal echoes was displayed by A-scan echography; spontaneous vertical movements of subretinal spikes indicating vascularity of the lesion could be observed on decreasing − 20 dB tissue sensitivity (Fig. 6); retrobulbar optic nerve widths were found normal in both eyes. Brain CT scan with contrast enhancement was negative and unable to show the lesion in the left eye. No photocoagulative treatment was performed.

No change was observed in visual acuity, fundus examination, fluorescein angiography or echography of the tumor during about a two-year follow-up.

Discussion

Fluorescein angiography is essential in the diagnosis of juxtapapillary capillary hemangiomas. B-scan echography is most sensitive for endophytic capillary angiomas; the high reflectivity of juxtapapillary capillary angiomas displayed by A-scan echography can differentiate them from other solid lesions such as choroidal melanomas which have low to medium acoustic reflectivity.[2] The lack of pigment and preretinal membrane in our two cases differentiates them from combined RPE and neuroretinal hamartoma which has also been reported to be a high reflective lesion without acoustic shadowing on standardized A-scan examination.[3] Moderate photocoagulation in multiple sessions between the juxtapapillary angioma and the macula should probably be used only in those tumors causing exudation and loss of vision prior to the development of macular detachment. In those tumors causing exudation and loss of central visual function moderately intense application of xenon or argon photocoagulation should be applied to selected portions of the tumor in multiple sessions.[1] The result of direct xenon photocoagulative treatment of the tumor in Case 1 was unsatisfactory.

Although juxtapapillary capillary angiomas show maximum growth during the second decade, they may occur in adulthood as in our Case 2. Since Gass *et al.*[1] and Schindler *et al.*[4] reported macular puckering following xenon photocoagulation of sessile juxtapapillary angiomas, no treatment was applied in our Case 2.

Curiously, both our patients with the rare juxtapapillary angioma came from a small area in Northern Italy.

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