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Bilateral CNV associated with optic nerve drusen treated with photodynamic therapy with verteporfin

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PURPOSE. To report a case of bilateral choroidal neovascularization (CNV) associated with optic nerve drusen (OND) treated with photodynamic therapy (PDT) with verteporfin.
METHODS. A 10-year-old girl with juxtapapillary CNV in the right eye and juxtapapillary and juxtafoveal CNV in the left eye associated with OND underwent PDT with verteporfin in both eyes.
RESULTS. Visual acuity increased from 20/160 to 20/25 in the right eye and from 20/1000 to 20/25 in the left eye after two sessions of PDT and 2 years of follow-up. CNV showed no leakage after two PDT sessions in both eyes and no recurrence was observed.
CONCLUSIONS. Subfoveal CNV is an uncommon complication of OND and excellent anatomic and functional results can be obtained with PDT. (Eur J Ophthalmol 2004; 14: 434-7)

KEY WORDS. Choroidal neovascularization, Photodynamic therapy, Optic nerve drusen, Verteporfin

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INTRODUCTION

Peripapillary and subfoveal choroidal neovascularization (CNV) has been described in association with isolated optic nerve drusen (OND) (1) and with POEMS syndrome (peripheral neuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and shin changes) plus optic disc drusen (2). CNV may cause severe and irreversible visual acuity loss in most cases (1-3).

Surgical treatment of subfoveal CNV has shown limited results (3). We describe a case of bilateral CNV associated with OND in a 10-year-old girl treated with photodynamic therapy (PDT).

Case report

A 10-year-old girl was referred to us with bilateral visual acuity decrease occurring 10 weeks previously, OND, and bilateral CNV (Fig. 1). Visual acuity in the right eye was 20/160 and in the left eye it was 20/1000. Ocular and systemic history were unremarkable. Serologic and hematologic study was negative, including for sarcoidosis and presumed ocular histoplasmosis syndrome. Ultrasound showed optic nerve hyperreflectivity with low gain in both eyes (Fig. 2). Fluorescein angiography revealed subfoveal hemorrhage in both eyes, juxtapapillary CNV in the right eye, and juxtapapillary and juxtafoveal CNV in the left eye (Fig. 1).

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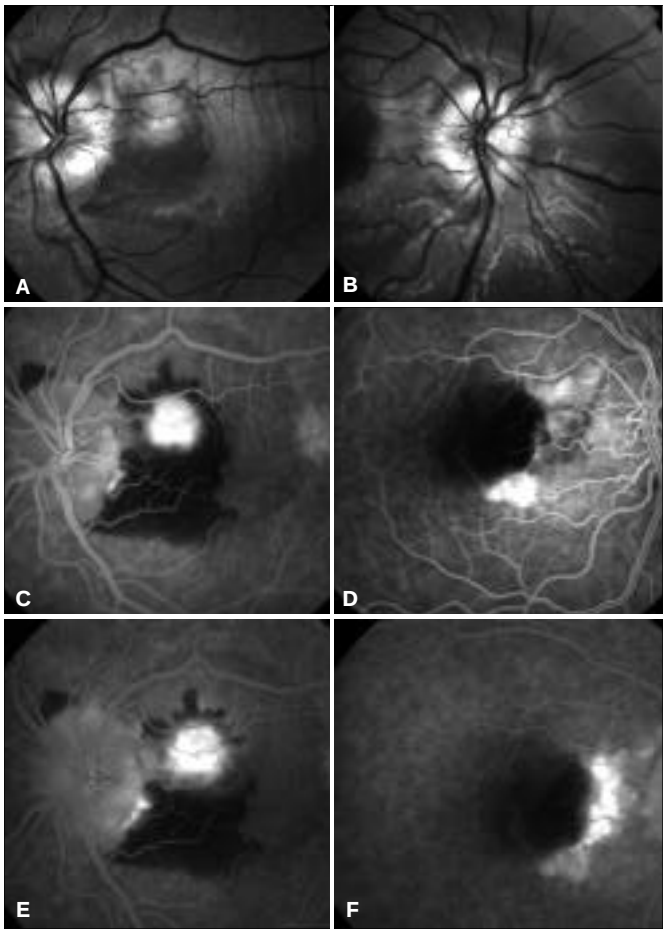

Fig. 1 - First visit: bilateral optic nerve drusen, subfoveal hemorrhage, and choroidal neovascularization (juxtapapillary in the left eye and juxtapapillary juxtafoveal in the right). Visual acuity was 20/160 in the left eye and 20/1000 in the right. (A, B) Red-free photography of left eye and right eye. (C, D) Early phases of fluorescein angiography (FA), left eye and right. (E, F) FA, left eye and right, late phases.

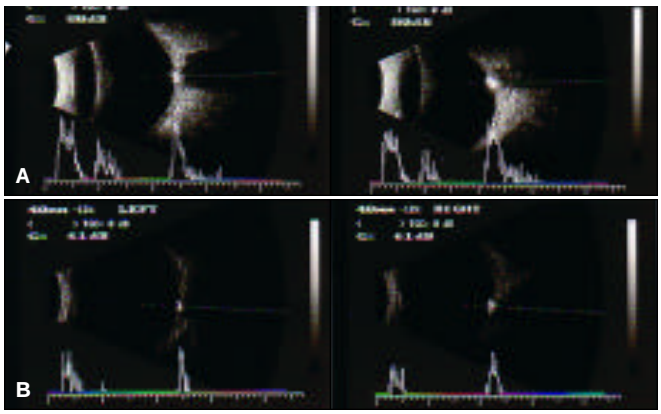

Fig. 2 - Ultrasound of left eye and right: with normal gain (A) and low gain (B). High reflectivity at the level of the papilla can be observed with normal and low gain.

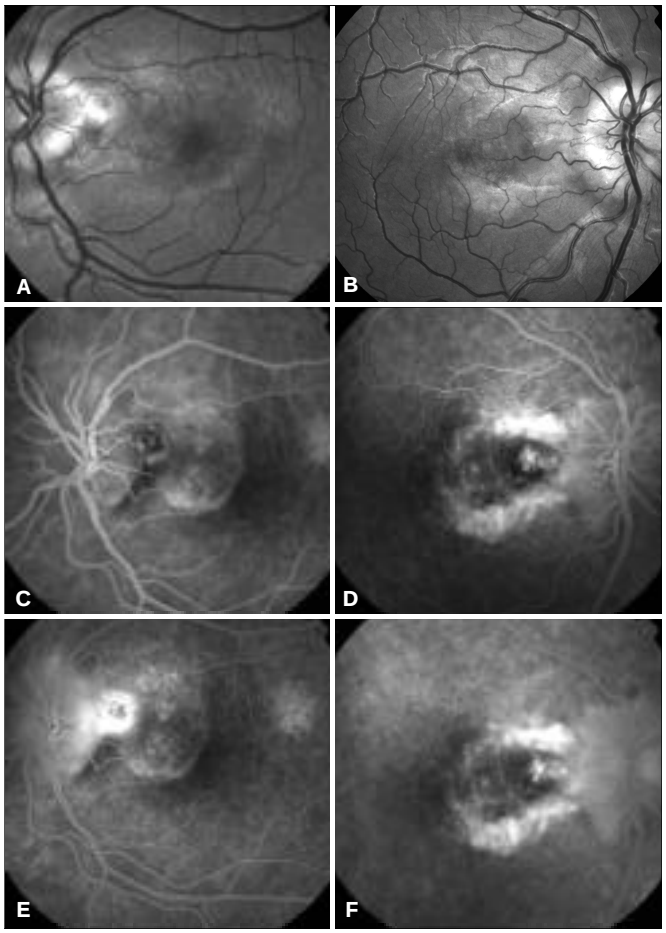

Fig. 3 - Five months later and after two photodynamic therapy sessions. (A, B) Red-free photography. No subretinal hemorrhages are visible. (C–F) Fluorescein angiography (FA): left eye shows diffuse subfoveal and sub-papillomacular bundle hyperfluorescence with no leakage. (D, F) Right eye still shows a juxtapapillary focal hyperfluorescence with questionable late leakage and a clear choroidal neovascularization size reduction (C, E). No additional photodynamic therapy was performed. Visual acuity improved to 20/32 in the left eye and 20/40 in the right.

PDT with standard parameters used in VIP (4) and TAP (5) studies was performed in both eyes in the same session.

Three months later, visual acuity was 20/40 in the right eye and 20/70 n the left and fluorescein angiography revealed some resolution of CNV but with persistence of leakage in both eyes. Bilateral PDT was repeated. Five months after the first treatment visual acuity was 20/32 in the right eye and 20/40 in the left and juxtapapillary fibrosis was observed

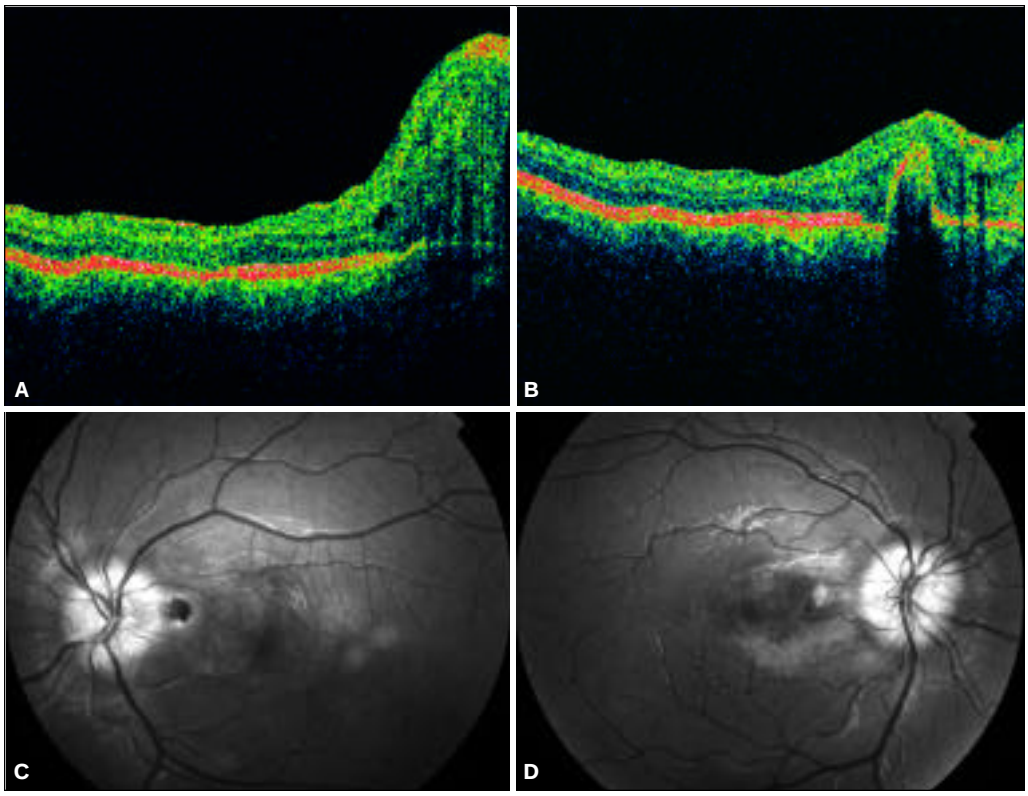

Fig. 4 - Optical coherence tomography and red-free photography at 2 years. A juxtapapillary scar is observed in the left (A, C) and right eye (B, D). Visual acuity improved to 20/25 in both eyes.

in both eyes (Fig. 3). No additional treatment was performed. The patient was observed at 3-month intervals with no recurrences for 2 years (Fig. 4). Final visual acuity was 20/25 in both eyes 24 months after the first treatment.

DISCUSSION

Several treatments have been applied to treat CNV in different pathologic conditions: laser photocoagulation, surgery, radiotherapy, transpupillary thermotherapy, and antiangiogenic and angiostatic drugs. VIP (4) and TAP (5) studies showed that PDT with verteporfin has efficacy and safety in some subtypes of subfoveal CNV. To our knowledge, no case reports of PDT with verteporfin for CNV associated with OND have been published.

VIP (4) and TAP (5) reported few cases of unexplained severe visual acuity loss following PDT with verteporfin. Bilateral treatments in the first session were not recommended. This unexplained visual acuity loss may occur after any treatment and not only after the first one. After explaining risks and benefits to the parents, bilateral PDT with verteporfin was performed in the same session, in the first and second visits, with success.

CNV related to inflammatory conditions, idiopathic CNV, or CNV associated with OND may show better surgical results when compared with age-related macular degeneration (4, 5). Surgery has been performed in all these situations, with relative success. However, loss of subfoveal pigment epithelium and photoreceptors with irreversible damage cannot be avoided. In one case reported with OND and bilateral CNV (3) surgery was performed for subfoveal CNV with visual acuity increasing from 0.05 to 0.3 and natural evolution was followed for the other eye with juxtapapillary CNV and stabilization of visual acuity at 0.3. In our case report, with PDT it was possible to stop the leakage and improve visual acuity without extensive anatomic damage, when compared with surgical excision of CNV.

CONCLUSIONS

Bilateral CNV associated with OND is a rare condition with no proven treatment. PDT with verteporfin can achieve excellent anatomic and functional results.

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