

Subretinal neovascularization associated with retinochoroidal coloboma

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PURPOSE. To report a case of subretinal neovascularization associated with retinochoroidal coloboma.

METHODS AND RESULTS. A 44-year-old female presented with metamorphopsia in her right eye for 4 weeks. Funduscopic examination revealed bilateral inferior retinochoroidal coloboma. Fluorescein angiography disclosed foci of subretinal neovascularization at the margin between the colobomatous defect and the normal-appearing retina. Five months later, multiple small areas of subretinal hemorrhages were noted. The hemorrhage was gradually absorbed. Six years after initial presentation, subretinal hemorrhage did not recur and her right VA was 0.2.

CONCLUSIONS. Ophthalmologists should be aware of this rare but important complication of retinochoroidal coloboma. (*Eur J Ophthalmol* 2005; 15: 815-7)

KEY WORDS. Retinochoroidal coloboma, Subretinal neovascularization, Photocoagulation

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INTRODUCTION

Retinochoroidal coloboma is a congenital developmental abnormality caused by incomplete closure of the ocular embryonic fissure.

Commonly accompanying ocular conditions include microphthalmia, cataract, and rhegmatogenous retinal detachment.

Subretinal neovascularization is a rare complication of choroidal coloboma with reports of only eight patients in the literature (Table I) (1-7).

We report an additional case of subretinal neovascularization associated with retinochoroidal coloboma.

Case report

A 44-year-old woman initially presented upon referral in March 1998 with metamorphopsia in her right eye for 4 weeks. On examination, visual acuity (VA) was 0.5 in the right and counts fingers in the left eye. Refractive errors were -2.25 in the right and -1.75 in the left eye. Intraocular

pressures were normal bilaterally. Slit lamp biomicroscopy disclosed bilateral inferonasal iris colobomas. Funduscopic examination revealed bilateral inferior retinochoroidal coloboma. The right defect extended to just below the fovea and did not involve the disc (Fig. 1a). The left defect involved the macula and disc. Fluorescein angiography (FA) disclosed foci of subretinal neovascularization at the margin between the colobomatous defect and the normal-appearing retina (Fig. 1b). Because of its juxtafoveal location and the patient's preference, laser photocoagulation to the choroidal neovascularization was not undertaken.

Five months later, multiple small areas of subretinal hemorrhages were noted. Her right VA decreased to 0.3. Fifteen months after initial presentation, serous retinal detachment of 2.5 disc diameters developed. Within the area of serous retinal detachment, a subretinal neovascular membrane of 1 disc diameter and surrounding subretinal hemorrhage were noted (Fig. 1c). Her right VA was 0.1. The hemorrhage was gradually absorbed and right VA improved to 0.2.

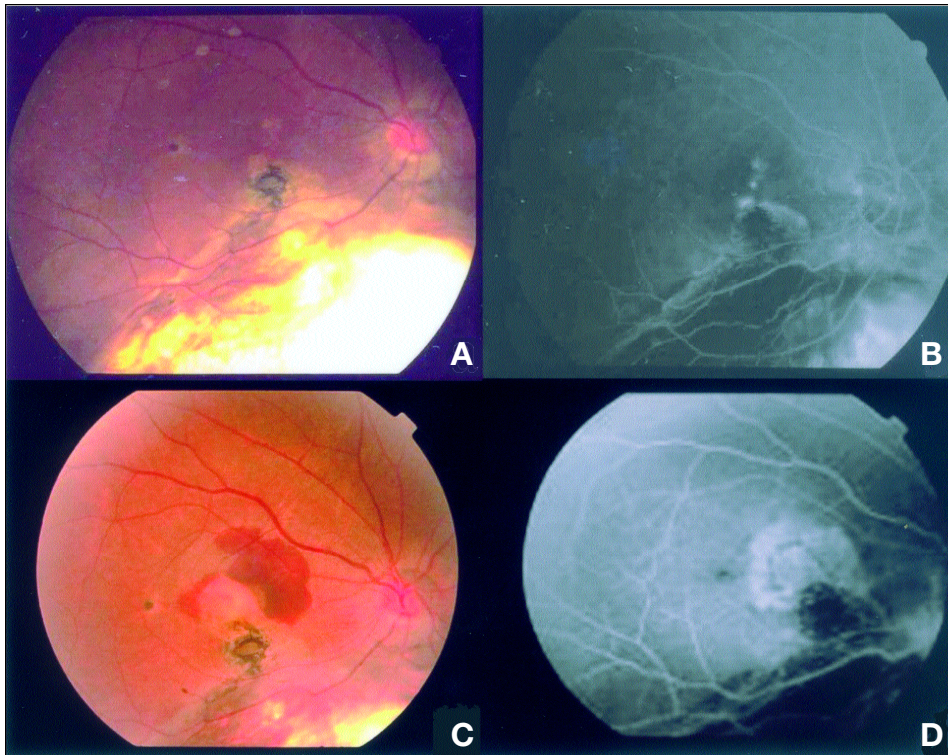


Fig. 1 - (A) Retinochoroidal coloboma in the right eye extended to just below the fovea but did not involve the disc. **(B)** Fluorescein angiography (FA) disclosed foci of subretinal neovascularization at the margin between colobomatous defect and the normal-appearing retina. **(C)** Within the serous retinal detachment, a subretinal neovascular membrane with surrounding hemorrhage was noted. **(D)** FA finding was indicative of tissue staining of disciform scar.

Three years after initial presentation, FA revealed hyperfluorescence of 1.5 disc diameters without progressive leakage, indicative of tissue staining of disciform scar (Fig. 1d). Subretinal hemorrhage did not recur and her right VA was 0.2 at her last visit in June 2004.

DISCUSSION

Subretinal neovascularization is a rare complication of retinochoroidal coloboma.

It was earlier reported in adults (1-4), but has subsequently been observed in infants (5, 7). Therefore, although degenerative aging changes may play an important role in the development of neovascularization in elderly patients (1, 3), the basis of the development of the disorder most likely can be ascribed to the disruption of the normal anatomy.

Schubert reported histologic sections (8) in eight choroidal colobomas and revealed that there was separation and reversal of direction in the periphery of outer retinal layer which became disorganized and connected to

TABLE I - REPORTED SUBRETINAL NEOVASCULARIZATION ASSOCIATED WITH CHOROIDAL COLOBOMA

Authors	Patient		Eye (R/L/B)	Photo-coagulation*	Visual acuity		Follow-up (mo)
	Age yr	Sex			Initial	Final	
Leff SR, et al (1985) (1)	65	F	R	N	20/400		
	70	F	R	N	20/70		
Steahly LP (1986) (2)	29	M	R	Y	20/200	20/30-2	33
Maberley AL, et al (1989) (3)	57	M	L	Y	20/400	20/50	
Rouland JF, Constantinides G (1991) (4)	26	F	R	N	CF	CF	10
Brodsky MC, et al (1991) (5)	1	F	R	Y		Fixation (+)	3
Gupta V, et al (1997) (6)	20	M	R	N	6/36		
Shaikh S, et al (2003) (7)	1	F	B	Y		Fixation (+)	10
Present case	44	F	R	N	0.5	0.2	74

*Application of laser photocoagulation to subretinal neovascularization
CF = Counts fingers

the retinal pigment epithelium, and that the choroid terminated peripheral to the point of reversal, both indicative of fragile mechanical barrier (*locus minoris resistentiae*) at the margin of the colobomas. The choroidal neovascularization that has been described at the margin of colobomas gains access to the subretinal space via the gap at this fragile barrier.

Although direct laser photocoagulation of the neovascularization was not offered in our patient because of its juxtafoveal location and the patient's preference, it may have possibly prevented decrease in vision (2, 3). However, the natural course of subretinal neovascularization associated with retinochoroidal coloboma remains to be elucidated.

Ophthalmologists should be aware of this rare but important complication of retinochoroidal coloboma, because timely laser photocoagulation was reported to be successful in selected patients (2, 3, 5, 7).

The authors have no proprietary interest in any aspect of the article.

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