

# Co-existent choroidal neovascular membrane and macular hole in pathologic myopia: a long follow-up clinical outcome and literature review

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## Abstract

• **Choroidal neovascularization (CNV) is an uncommon complication associated with a macular hole. In this case report of a rare condition, we present a pathologic myopia patient with a co-existent macular hole and choroidal neovascular membrane. The patient was treated with photodynamic therapy for CNV, and then vitreous surgery for the retinal detachment and macular hole. At the end of 4 years follow-up, her visual acuity was improved to 0.1 while the macular hole remained open. Optical coherence tomography is a useful inspection method of the diagnosis of CNV and macular hole.**

• **KEYWORDS:** choroidal neovascularization; macular hole; pathologic myopia

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## INTRODUCTION

Pathologic myopia (PM) is characterized by an axial length exceeding a certain threshold (typically 26.5mm, corresponding to a refractive error of at least -6.0 diopters)<sup>[1]</sup> accompanied by certain pathologic changes. Myopic

maculopathy comprises features of progressive macular degeneration, choroidal neovascularization (CNV) formation, along with macular hole, posterior staphyloma and retinoschisis. The co-existence of a macular hole and CNV is an extremely rare occurrence. The pathogenesis, clinical presentation, diagnosis and treatment of choroidal neovascular membrane and macular hole in pathologic myopia are reviewed.

## TYPICAL CASE

A 59-year-old woman complained of decreased vision of her left eye for several days. Her best corrected visual acuity (BCVA) was 0.2 in left eye tested with decimal visual acuity chart. Both eyes had pathologic myopia (-12D). Her right eye had a macular hole, and she received an Argon focal photocoagulation in the macula 14 years ago. Her central vision of the right eye was decreased to 0.02. There was a large atrophy area in the macula. When her left eye had first developed a full-thickness macular hole, she was not treated with laser, yet her BCVA remained stable for almost 5 years. Further visual decline was noted because of newly developed classic juxtafoveal CNV (Figure 1A) which was confirmed by optical coherence tomography (OCT) (Figure 1B) and fundus fluorescein angiography (Figure 1C, D). After 14d, retinal detachment was observed in the macular area (Figure 1E, F). The temporal and inferior quadrant was involved in quickly (Figure 1G). A linear OCT scan showed the full-thickness macular hole with a choroidal neovascular membrane and retinal detachment (Figure 1H). Her visual acuity of the left eye declined to finger counting/20cm. The CNV was treated with photodynamic therapy prior to macular hole surgery. After 5d, the patient underwent a pars plana vitrectomy combined with membrane separation, removal of the internal limiting membrane, air fluid exchange, and injection with silicone oil. Three months after the surgery, the patient's BCVA of left eye improved to 0.1. A dilated fundusoscopic examination showed a stable CNV with a flattening of the surrounding pigment epithelium. OCT examination confirmed a resolution of the neuroepithelium detachment. After 10 months, the silicone

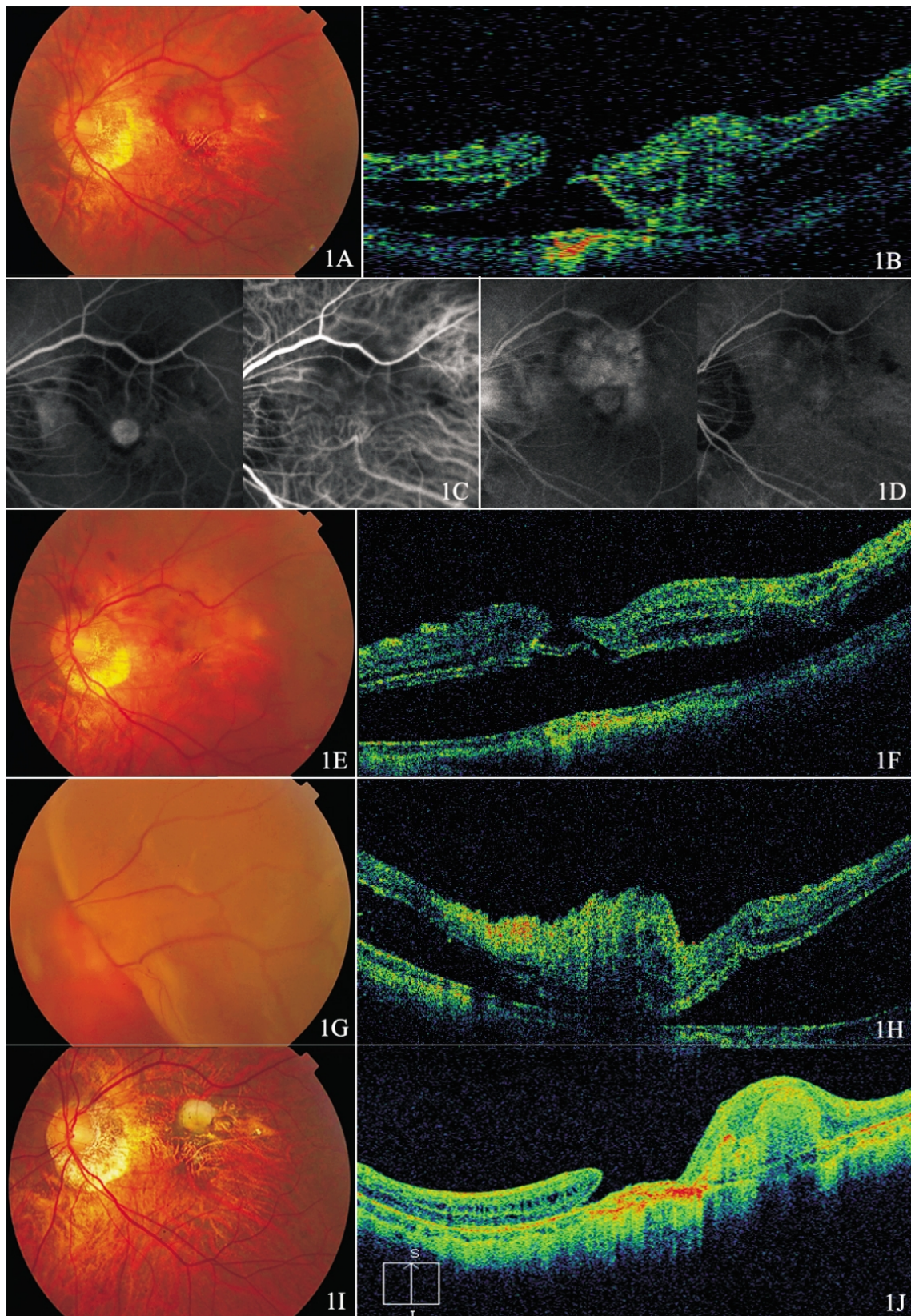


Figure 1 Fundus photograph, fundus fluorescein angiography and OCT results of the patient.

oil was removed. To date, the retina has remained re-attached for 4 years (Figure 1I). Her BCVA and CNV is stable, while the macular hole is still open (Figure 1J).

#### DISCUSSION

Myopic maculopathy is characterised by the presence of one or more of the following changes: posterior staphyloma,

lacquer cracks and myopic CNV, macular hole and chorioretinal atrophy in the posterior fundus. This report discusses a case of a macular hole in which no retinal detachment developed during the long follow-up period of a 59-year-old Chinese female with high myopia. When an active CNV occurred, retinal detachment developed within

two weeks.

**Pathogenesis** Pathologic myopia is associated with a progressive elongation of the globe of the eye, which results in various funduscopic changes in the macular area [2]. Chen *et al* [3] reported that the type of high myopic maculopathy had a statistically significant association with refractive error. In a previous study, Shimada *et al* [4] demonstrated that the incidence of macular holes was relatively high (14.0%) in highly myopic eyes at the atrophic stage of CNV. A histopathologic study has shown that there is a loss of the retinal pigment epithelium (RPE) layer and the outer retinal layer in the area of the chorioretinal atrophy in myopic eyes at the atrophic stage of myopic CNV [2].

The causative factors of detachment in macular hole were refractive error, myopic chorioretinal change and posterior staphyloma. Pathologic myopic eyes with a macular hole are almost always associated with retinal detachment [5]. Our patient was interesting because no retinal detachment was observed, even though the patient did have a macular hole with pathologic myopia and posterior staphyloma. The patient subsequently had a rapidly appearing serous retinal detachment that maybe related to the permeability and activity of the CNV. Fluid leakage and inflammatory mediators, such as the vascular endothelial growth factor (VEGF) from the CNV itself, or damage of the retinal pigmented epithelium might have played a role in the retinal detachment.

**Clinical Presentation** CNV occurs in approximately 5% of eyes with pathologic myopia and may lead to central vision loss [2]. A study of case records from 218 patients (325 eyes) with myopic fundus changes showed that approximately 10% of eyes developed CNV during an average of almost 11 years of follow-up [6]. CNV, a common cause of vision loss in pathologic myopia, may often cause irreversible deterioration of central vision. The precise pathogenesis of CNV in eyes with pathologic myopia is unclear, although thinning of the choroid and stretching of retinal tissue appear to be involved. Until recently, treatment options for CNV were limited. Laser photocoagulation and photodynamic therapy (PDT) with verteporfin have shown beneficial effects compared with no treatment in selected cases of CNV secondary to pathologic myopia [7]. Anti-VEGF becomes a new treatment tendency. The recent introduction of VEGF inhibitors has revolutionized the management of CNV secondary to pathological myopia. Intravitreal ranibizumab, given as monthly injections or as 3 monthly injections followed by as needed dosing, showed a promising efficacy and safety profile in the treatment of CNV due to causes other than AMD [8]. Intravitreal bevacizumab injection is another

beneficial treatment for CNV associated with pathologic myopia [9,10].

A macular hole retinal detachment almost always develops in eyes with a posterior staphyloma. Posterior staphyloma rather than anteroposterior vitreomacular traction may contribute to the development of retinal detachment associated with a macular hole in severely myopic eyes [11]. Various surgical procedures, including vitrectomy and macular scleral buckling, are used to repair this disorder, although the success rate is currently not high [12-14].

**Diagnosis** An fundus ophthalmoscopy and fundus fluorescein angiography and OCT were used to access the clinical finding of CNV and the macular hole. Fluorescein angiography is the established method for evaluating whether any therapy is indicated for CNV due to pathologic myopia. Most CNV secondary to pathologic myopia exhibits minimal leakage, perhaps because the source of the blood supply, the choriocapillaris, is severely reduced by stretching of the ocular tissues. OCT is a non-invasive tool for the quantitative assessment of retinal thickness and has become the standard of care in the management of myopic CNV, as in other macular diseases. It is well known that classic CNVs located above the RPE cell layer (Type 2 CNV) are better defined using OCT [15], and thus, OCT is particularly useful in myopic CNV because the vast majority of myopic CNVs are classic in nature. In the active stage, myopic CNV appears as highly reflective dome shaped elevation above the band corresponding to the RPE with negligible subretinal fluid accumulation. OCT may be helpful to identify co-existing pathologies related to myopia, such as macular hole and retinoschisis. From our case, we suggest that patients with co-existent macular hole and CNV need to be evaluated every week in the first month and every three months in the late stage.

**Treatment** The management of these two lesions within the same eye is a therapeutic dilemma. Only a handful of cases with a macular hole and CNV in the same eye have been reported [16,17]. Moreover, the previous reports have indicated different visual results [18,19]. The active CNV should be treated so as to halt its activity. We chose the treatment for the CNV with photodynamic therapy because bevacizumab injection was off label and ranibizumab was unavailable at that time. When retinal detachment occurs, pars plana vitrectomy should be performed. During a period of at least 4 years of follow-up in this series, the retina and visual acuity remained in a stable condition. For these patients with cases of a complicated pathology of myopia maculopathy patients, it is necessary that individualized treatments should be worked out.

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