Idiopathic Unilateral Choroidal Neovascularization in a Young Female

Brian Nguyen, D.O., Mary Gao, D.O., Inna Bondira, D.O., Vikisha Hazariwala, D.O.; Kamal Gupta, M.D.

Corewell (Beaumont) Ophthalmology ■ Corewell Health – Taylor, MI

INTRODUCTION

Choroidal neovascularization (CNV) is a significant cause of visual disability and morbidity in older individuals with age-related macular degeneration (ARMD). However, it can also occur in individuals younger than 50 years of age with certain predisposing conditions, such as pathological myopia, angioid streaks, inflammatory chorioretinopathies (e.g. white dot syndromes, ocular histoplasmosis), choroidal neoplasms, traumatic choroidal rupture, and optic nerve head abnormalities.[1, 2] Even still, a proportion of young patients with CNV will not present with any of the aforementioned risk factors, constituting the idiopathic subtype of CNV (ICNV). Typically, these membranes are found unilaterally and visual prognosis, which is largely dependent on the size of the membrane, is often more favorable than CNV resulting from ARMD and pathologic myopia.[3, 4]

ICNV in the younger population represents a minority of cases not often described in the literature with unknown incidence. Various methods of treating unilateral ICNV have been studied, the most promising of which include photodynamic therapy (PDT) and injection of intravitreal anti-vascular endothelial growth factor (VEGF) agents.[5, 6, 7] Both procedures are considered to be relatively safe and well-tolerated in the management of ICNV. We report an idiopathic unilateral, subfoveal ICNV in a 22-year-old female who ultimately responded favorably to a series of intravitreal bevacizumab injections over a period of five months.

CASE REPORT

We report a case of a 22-year-old Caucasian female who presented with a sudden onset of blurred vision in her right eye (OD) one week prior to presentationt. She also endorsed metamorphopsias, but denied eye pain, flashes of light or floaters. There was no history of ocular trauma. Medical and ocular history was significant for COVID-19 respiratory infection one month prior to presentation and low myopia, respectively. Examination findings from the first visit are provided in Table 2.

OCT of the macula (Figure 1), ultra wide-field fundus photography, and fluorescein angiography (Figure 2) were performed and revealed leakage at the macula consistent with choroidal neovascularization. Following the imaging studies, the patient was immediately started on intravitreal injections with bevacizumab and maintained at six-week intervals until resolution of macular edema was noted. Table 1 and Figure 1A-C demonstrate the patient's response throughout her treatment course. She has thus far received four intravitreal injections with resolution of macular edema, and will be followed closely over the coming months and years due to the risk of recurrence.

	BCVA OD
Visit 1: 10/31/22	20/60
Visit 2: 11/3/22 * First intravitreal bevacizumab injection	20/400-
Visit 3: 11/10/22	20/70
Visit 4: 12/19/22	20/30
Visit 5: 3/13/23	20/20

Table 1. Best corrected visual acuity (BCVA) of the affected eye.

Visit 1	Right eye	Left eye
Eyelids, Adnexa	WNL	WNL
Conjunctiva, Sclera	White and quiet	White and quiet
Cornea	Clear	Clear
Anterior chamber	Deep and quiet	Deep and quiet
Iris	Flat and round, no synechiae	Flat and round, no synechiae
Lens	Clear	Clear
Vitreous	Clear	Clear
Optic nerve	Pink with sharp margins, 0.3	Pink with sharp margins, 0.3
Macula	Subfoveal elevation and subretinal hemorrhage	Flat and attached
Vessels	Normal caliber	Normal caliber
Periphery	Flat and attached with 360 degree scleral depression	Flat and attached with 360 degree scleral depression

Table 2. Physical examination findings on presentation

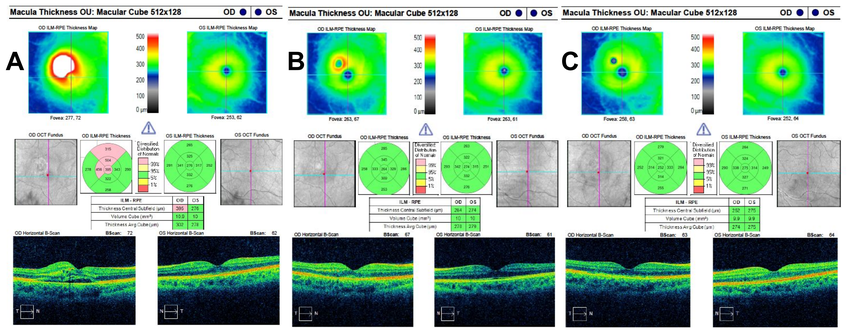

Figure 1. A). OCT on the day of presentation showing CNV under the retinal pigment epithelium (RPE) and intra- and sub-retinal fluid OD. B, C). OCT 1 week and 6.5 weeks after intravitreal bevacizumab injection, showing marked improvement in intra- and sub-retinal fluid OD.

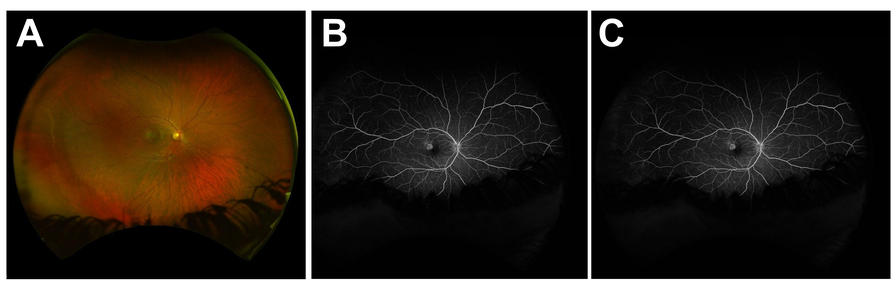

Figure 2. Images taken prior to the first intravitreal injection. A). Fundus photo OD showing macular edema and subretinal hemorrhage. B,C). Early (B) and late (C) phase of fluorescein angiography (FA) showing a well-demarcated area of hyperfluorescence with leakage indicative of CNV.

DISCUSSION

Idiopathic CNV is a diagnosis of exclusion after evaluating well-established causes.[1,2] All of these causes had been ruled out in our patient, including pathologic myopia, trauma, angioid streaks, and ocular histoplasmosis. Our leading differential diagnosis for this patient was CNV secondary to central serous chorioretinopathy (CSCR), however, fluorescein angiography demonstrated fluorescein leakage without a focal source, rather than pooling which is seen in CSCR.

Our 22-year-old female patient demonstrated many typical CNV findings in the absence of any known risk factors, which led us to the diagnosis of ICNV. One prior case report suggests that ICNV in young adults, particularly females, may be a precursor or early manifestation of an inflammatory chorioretinopathy[2]. The inflammatory chorioretinopathies in these patients developed CNV in either the ipsilateral or contralateral eye.[2]

Treatment is currently focused on using a series of intravitreal anti-VEGF injections. Other considerations should be given to photodynamic therapy (PDT) as an alternative treatment for subfoveal CNV secondary to idiopathic or inflammatory causes.[8] Overall, anti-VEGF therapy has been found to be superior to PDT for ICNV, and thus serves as the mainstay of treatment in ICNV patients.[5,7]

Our case report represents another great example of the effectiveness of anti-VEGF treatment for ICNV in young adults. Special care should be taken in using anti-VEGF therapy in female patients where pregnancy is of concern, as anti-VEGF medicines are designated as Category C by the FDA[9] due to the risk of abnormalities in embryogenesis leading to fetal loss[10]. Despite this, there is still limited evidence of the maternal-fetal transplacental transmission.[9,11] Due to these concerns, a discussion on birth control is warranted with females of reproductive age who are undergoing anti-VEGF therapy.

CONCLUSION

While our patient responded favorably to anti-VEGF therapy, further investigation into the natural disease course and pathophysiology of ICNV may help elucidate visual prognosis and patient-directed treatments to further our understanding of this complex disease entity.

REFERENCES

1. Cohen SY, Laroche A, Leguen Y, Soubrane G, Coscas GJ. Etiology of choroidal neovascularization in young patients. *Ophthalmology.* 1996;103:1241–4.
2. Machida S, Fujiwara T, Murai K-I, et al. Idiopathic choroidal neovascularization as an early manifestation of inflammatory chorioretinal diseases. *Retina*. 2008;28(5):703–710.
3. Ho AC, Yannuzzi LA, Pisicano K, DeRosa J. The natural history of idiopathic subfoveal choroidal neovascularization. *Ophthalmology.* 1995;102:782–9.
4. Lindblom B, Andersson T. The prognosis of idiopathic choroidal neovascularization in persons younger than 50 years of age. *Ophthalmology.* 1998;105:1816–20.
5. Kang HM, Koh HJ. Intravitreal anti– vascular endothelial growth factor therapy versus photodynamic therapy for idiopathic choroidal neovascularization. Am J Ophthalmol. 2013; 155(4):713–719.
6. Spaide RF, Martin ML, Slakter J, Yannuzzi LA, Sorenson J, Guyer DR, et al. Treatment of idiopathic subfoveal choroidal neovascular lesions using photodynamic therapy with verteporfin. *Am J Ophthalmol.* 2002;134:62–8..
7. Cakir M, Cekiç O, Yilmaz OF. Intravitreal bevacizumab for idiopathic choroidal neovascularization. *J AAPOS*. 2009;13:296–8.
8. Rogers AH, Duker JS, Nichols N, Baker BJ. Photodynamic therapy of idiopathic and inflammatory choroidal neovascularization in young adults. Ophthalmology. 2003 Jul;110(7):1315-20. doi: 10.1016/S0161-6420(03)00466-4. PMID: 12867384.
9. Galazios G., Papazoglou D., Tsikouras P., and Kolios G. Vascular endothelial growth factor gene polymorphisms and pregnancy. J. Matern. Fetal Neonatal. Med. 22:371–378, 2009
10. Naderan, M., Sabzevary, M., Rezaii, K. et al. Intravitreal anti-vascular endothelial growth factor medications during pregnancy: current perspective. Int Ophthalmol 41, 743–751 (2021). https://doi.org/10.1007/s10792-020-01610-2
11. Sullivan L, Kelly SP, Glenn A, Williams CP, McKibbin M. Intravitreal bevacizumab injection in unrecognised early pregnancy. Eye (Lond). 2014 Apr;28(4):492-4. doi: 10.1038/eye.2013.311. Epub 2014 Jan 17. PMID: 24434664; PMCID: PMC3983635.