Choroidal neovascularization secondary to choroidal nevus simulating an inflammatory lesion

Samuray Tuncer, Ilknur Tugal-Tutkun

Choroidal nevi are the most common benign pigmented lesions of the fundus. Choroidal neovascularization is a rare complication of choroidal nevi. We report herein a young patient managed successfully with intravitreal bevacizumab injections for juxtapapillary choroidal neovascularization secondary to choroidal nevus simulating an inflammatory lesion.

Key words: Bevacizumab, choroid, choroidal neovascularization, choroidal nevus, nevus

Choroidal nevi are the most common benign pigmented lesions of the fundus. Prevelance estimates range from 4% to 8% of the population.[1] Secondary retinal and retinal pigment epithelium (RPE) changes include drusen, atrophy and/or hyperplasia of RPE, fibrous metaplasia, orange pigment, RPE detachment, and subretinal fluid. Choroidal neovascularization (CNV) is a rare complication of choroidal nevi. Li and coauthors reported CNV in less than 1% of eyes with giant choroidal nevi.[2] We report herein a young patient managed successfully with intravitreal bevacizumab injection for juxtapapillary CNV secondary to choroidal nevus simulating an inflammatory lesion.

Case Report

A 16-year-old girl was referred to our clinic with a diagnosis of neuroretinitis or juxtapapillary chorioretinitis. The patient had a 3-week history of decreased vision and metamorphopsia in her left eye. Her medical and family history were unremarkable. There was no history of trauma or high myopia. Before referral, systemic work-up for tuberculosis, sarcoidosis, syphilis, Lyme disease, brucellosis, and toxoplasmosis was negative. At our initial visit, best-corrected visual acuity was 20/20 in the right eye and counting fingers in the left eye. There was a mild relative afferent pupillary defect in the left eye. Biomicroscopic examination was within normal limits. There was no cellular reaction in the anterior chamber or vitreous cavity. The intraocular pressure was 16 mmHg in both eyes. Fundus examination of the left eye revealed a juxtapapillary, elevated, white chorioretinal lesion consistent with CNV that was associated with exudation and retinal edema [Fig. 1a]. The juxtapapillary choroidal nevus underlying CNV was not visible with color fundus photography [Fig. 1a], but we were able to detect it with the brightest illumination of the indirect ophthalmoscope. Fundus fluorescein angiography (FFA) demonstrated extrafoveal, juxtapapillary CNV, and late diffuse hyperfluorescence superior to this lesion. Optic disc was normal in all phases of the angiography [Fig. 1b-c]. Optic coherence tomography (OCT) confirmed CNV with overlying intraretinal cysts and minimal subretinal fluid. A diagnosis of juxtapapillary CNV associated with a choroidal nevus was made. After a written informed consent was obtained, three consecutive monthly intravitreal bevacizumab (1.25 mg/0.05 ml) injections were performed. After treatment, best-corrected visual acuity improved to 20/50. CNV appeared inactive and smaller in size with complete resolution of exudation exposing an underlying choroidal nevus with 6 × 6 × 2.6 mm in dimensions [Fig. 1d]. OCT demonstrated gradual resolution of subretinal fluid and intraretinal cysts [Fig. 2]. FFA revealed burnt-out fibrotic CNV with late staining and juxtapapillary hypofluorescence due to underlying choroidal nevus [Fig. 1e]. The lesion was still inactive and final visual acuity was stable throughout 2 years of follow-up.

Discussion

The case presented herein shows the rare association of CNV with a choroidal nevus in a 16-year-old girl. In a young patient with CNV, inherited and acquired conditions such as angioid streaks, high myopia, trauma, choroidal tumors, familial macular dystrophies, and inflammatory retinochoroidopathies should be considered in the differential diagnosis.

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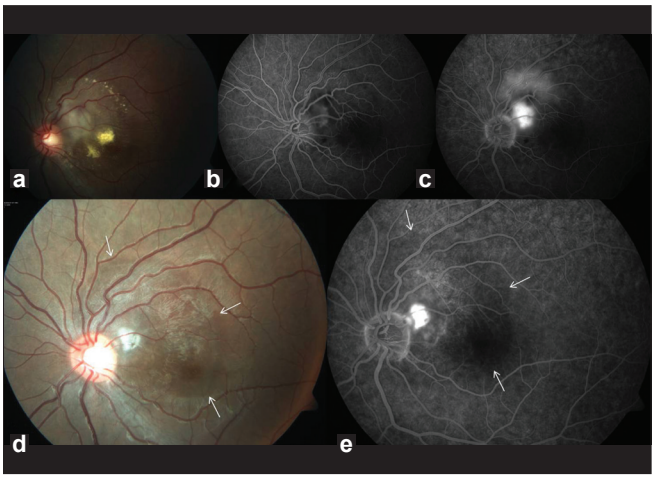

Figure 1: Before treatment: (a) juxtapapillary, elevated, white chorioretinal lesion with exudation, (b) FFA (early) reveals faint juxtapapillary hyperfluorescence of CNV and pin-point hyperfluorescent points superior to this lesion, (c) FFA (late) shows juxtapapillary CNV and diffuse leakage. After treatment: (d) complete resolution of exudation exposing the underlying choroidal nevus (arrows), CNV appears inactive and smaller in size, (e) FFA reveals burnt-out fibrotic CNV with late staining and ill-defined juxtapapillary hypofluorescence due to underlying choroidal nevus (arrows)

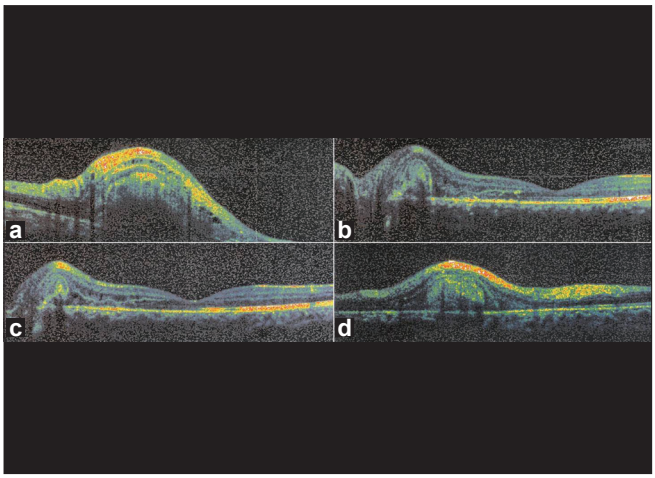

Figure 2: OCT shows gradual regression of CNV and complete resolution of intraretinal edema and subretinal fluid with restoration of the retinal architecture, (a) CNV associated with minimal subretinal fluid and intraretinal cystic edema (initial visit), (b) after first injection, (c) after second injection, and (d) after third injection

Juxtapapillary CNV may be mistaken for neuroretinitis or chorioretinitis. The absence of optic disc edema, vitritis, macular star, or optic disc hyperfluorescence on FFA helped us rule out an inflammatory lesion. Our case shows that an underlying choroidal nevus may not be readily perceivable as a cause of CNV when there is prominent exudation from the CNV. Most of the ophthalmologists would prefer to perform an indocyanine green angiography to detect the underlying pathology in the presence of a CNV in a young patient. However, we were able to detect the underlying choroidal nevus with indirect ophthalmoscopy and managed successfully with intravitreal bevacizumab injections.

Choroidal nevus-related CNV is a rare entity and may be either extrafoveal or subfoveal. Treatment options include laser photocoagulation, transpupillary thermotherapy, and photodynamic therapy.[1,3,4] Laser photocoagulation can be used in extrafoveal CNV, but it destroys the overlying retina and the pigment epithelium.[3] Visually significant scotomas may develop when laser is applied for subfoveal CNV and lesions located in the papillomacular bundle due to irreversible damage to nerve fiber layer. Transpupillary thermotherapy may also be an option for juxtafoveal and extrafoveal CNV. Possible thermal injury to underlying retinal and retinal pigment epithelial structures may lead to atrophic changes in larger areas that may threaten the visual acuity.[4] Photodynamic therapy is preferred in the management of subfoveal CNV. Vascular complications including optic nerve ischemia and retinal vessel occlusions were reported in juxtapapillary choroidal hemangiomas managed with photodynamic therapy.[5] Considering these potential complications, we preferred intravitreal bevacizumab injection for the management of juxtafoveal CNV which was found to be secondary to a choroidal nevus. We did not observe any adverse event and we achieved both anatomical and functional improvement with this treatment modality.

In conclusion, an underlying choroidal nevus should be considered in the differential diagnosis of CNV in young patients. To our knowledge, our patient represents the first case of choroidal nevus-related CNV successfully treated with intravitreal bevacizumab injections.

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