

Bilateral circumscribed choroidal hemangioma: a case report

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Dear Editor,

Bilateral circumscribed choroidal hemangioma (CCH) is mostly associated with Sturge-Weber syndrome and rarely occurs in healthy individuals. So, we present the case about bilateral CCH that occurred in a healthy person and to evaluate the efficacy of 0.05 mL of anti-vascular endothelial growth factor (anti-VEGF) medication and laser photocoagulation in the treatment of CCH. We obtained the written informed consent from the patient, and this case study was in accordance with the tenets of the Declaration of Helsinki.

A 21-year-old young man with no past medical history presented to our department with a chief complaint of one week of decreased left eye vision. The physical examination showed that his visual acuity was 0.02 in the left eye and 0.08 in the right eye. There were no obvious abnormalities in the

anterior segment and vitreous of both eyes under slit lamp. In the right eye, the fundus retina was flat, and a round-shaped lesion with a clear border of about 3.0 papilla diameter (PD) was seen in the temporal peripheral retina, with no hemorrhage or exudation on the surface of the lesion; in the left eye, a greyish-white lesion of 4.5 PD was seen in the fundus of the left eye, with a slight elevation of the retina around the lesion, and the retina of the macular region was edematous and thickened (Figure 1A).

Accessory examinations were performed to further explore the disease. B-ultrasound showed a solid hemispherical mass lesion in the left eye (Figure 1B). Optical coherence tomography (OCT) showed that the neuroepithelial layer of the macular region of the left eye was detached (Figure 1C). Fluorescein angiography (FFA) showed that focal mottled fluorescence in the temporal side of the right eye, choroidal space-occupying lesion in the left eye, which formed a discoid fluorescent pool accumulation area in the lower part, involving the macular area. Indocyanine green angiography (ICGA) showed that choroidal vascular clusters can be seen within the lesion during early imaging, and as the imaging time prolongs, the fluorescence leakage within the lesion appears as a strong fluorescence lesion, while there was flushing phenomenon in the later stage (Figure 1D and 1E). Furthermore, magnetic resonance imaging (MRI) of the head and neck showed that a small protrusion was seen at the lower edge of the ophthalmic segment of the left internal carotid artery, suggesting a small aneurysm, with no other obvious abnormalities. The patient's chest computed tomography (CT) and blood tests did not show any significant abnormality.

All evidences suggested that it might be bilateral CCH with plasma retinal detachment in the left eye, so we performed retinal laser photocoagulation in the right eye and 0.05 mL of conbercept in the vitreous cavity of the left eye. Subsequently, the patient came for follow-up every month, and according to the patient's condition, retinal laser photocoagulation in the focal area of the left eye and intravitreal injection were performed for two times. The best-corrected visual acuity (BCVA) of the left eye was 0.2 and a significant improvement was observed in retinal structure at 2mo and 3mo after surgery

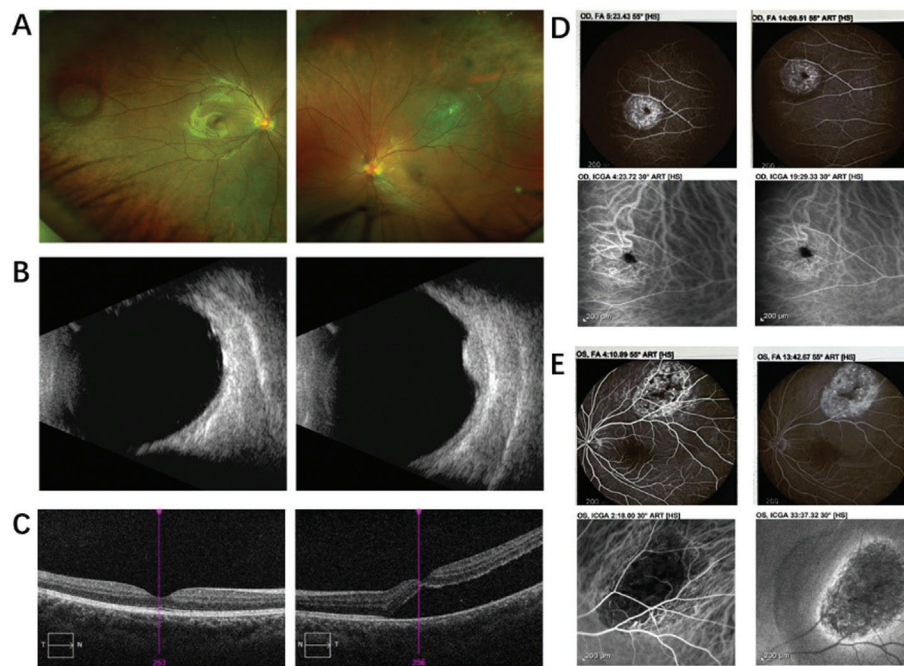


Figure 1 Images of case before treatment A: Scanning fundus laser photography of both eyes. B: B-ultrasound: No obvious abnormal echo in the right eye, and a hemispherical moderate uniform echo can be seen in the left eye. C: OCT: The shape of the retina in the macular area of the right eye was normal at presentation, and the neuroepithelial layer of the macular region of the left eye was detached. D: FFA of right eye showed irregular high fluorescence mixed with low fluorescence in the lesions. ICGA showed that round mottled strong fluorescence of the periorbic omentum of the temporal side of the right eye. E: Abnormal linear and granular high fluorescence within the lesion can be seen in the early left eye FFA while discoid fluorescence accumulation can be seen below the lesion in the late stage. ICGA of left eye showed that persistent high fluorescence was present in the early stage of the lesion, while low fluorescence was present in the late stage (flushing phenomenon) with a high fluorescence ring can be seen around the lesion. OCT: Optical coherence tomography; FFA: Fluorescein angiography; ICGA: Indocyanine green angiography.

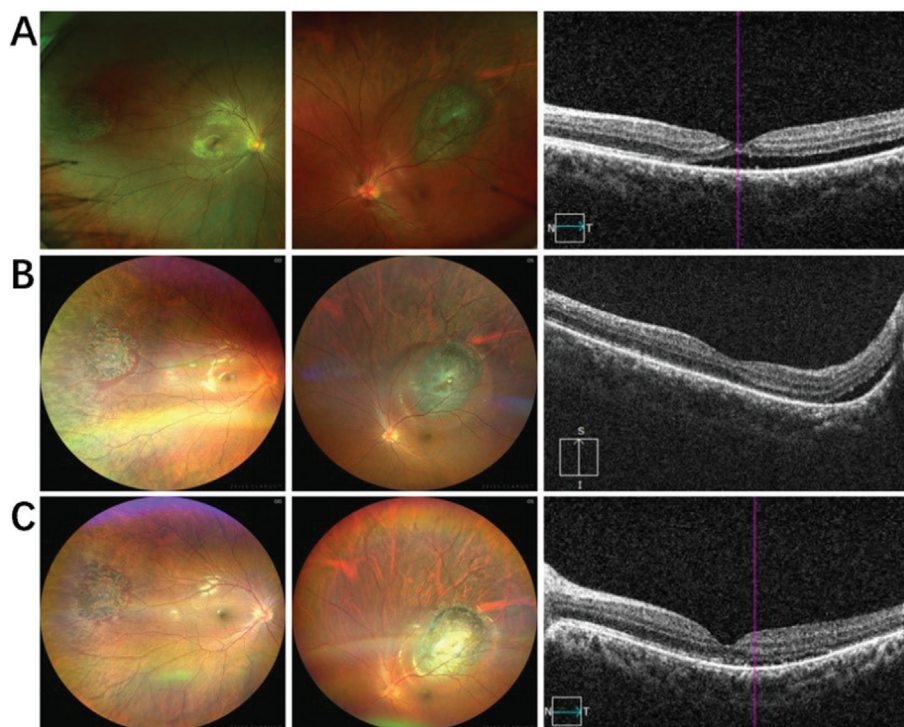


Figure 2 Images of case after treatment Two (A), 3 (B), and 6mo (C) after surgery, subretinal fluid in the left eye gradually absorbed and completely disappeared at the sixth month while the condition of the right eye was keeping stable and unchanged.

(Figure 2A and 2B). Six months after the surgery, the BCVA of the left eye had not improved further, but subretinal fluid was completely absorbed with the thickness of the retina mostly returned to normal (Figure 2C). The BCVA of the right eye

was 1.0, the retina in the right eye lesion area was flat and maintain stability for the next six months.

CCH is a congenital benign hemangioma that is easily overlooked or misdiagnosed as a malignant lesion in routine ophthalmic examinations. It usually appears as a well-defined orange-red mass in the posterior pole of one eye, while has a grey-green hue in scanning fundus laser photography^[1], that is why it is easily misdiagnosed as choroidal melanoma. MRI and ultrasound are helpful in differentiating CCH from choroidal melanoma and metastases^[2]. As hemangioma leakage increases, patients may come to seek medical attention due to symptoms of visual loss caused by subretinal fluid accumulation or macular edema^[3]. It needs to be distinguished from central serous chorioretinopathy (CSC), which also manifests as a clear fluid accumulation under the retinal neuroepithelium in the posterior pole-like circular area and shows a fluid cavity between the retinal neuroepithelium and the pigment epithelium in the macular area in OCT^[4]. FFA and ICGA is the most diagnostic test for choroidal hemangiomas, especially ICGA, with the “dye washout phenomenon” formed by the high fluorescence in the early stages and the low fluorescence in the late stages being the typical features of choroidal hemangiomas^[5]. Treatment has evolved from laser photocoagulation to photodynamic therapy (PDT), external beam radiotherapy and intravitreal injection of anti-VEGF^[6].

In this case, we diagnosed the patient with CCH in both eyes combined with plasma retinal detachment involving the macula in the left eye. Given the rarity of bilateral CCH, a single ophthalmological examination is prone to misdiagnosis and underdiagnosis, multimodal imaging is particularly important for the diagnosis and differential diagnosis of CCH^[7]. Ultrasound shows a dome-shaped dense echogenic mass, MRI demonstrates hyperintensity on T1-weighted images and isointensity on T2-weighted images when compared with vitreous, as well as the washout phenomenon in ICGA, may be suggestive of the diagnosis of CCH. Meanwhile, the presentation of the patient’s right eye was relatively atypical, with no significant abnormalities on MRI or ultrasound, so a definitive diagnosis with the help of FFA and ICGA was extremely necessary. In the development of the treatment plan, due to the high cost of PDT, shortage of drugs, and postoperative complications related to PDT, such as retinal pigment epithelial atrophy and hyperplasia, and subretinal fibrosis, *etc*^[8], and taking into account that the purpose of CCH is not to eliminate the hemangioma, but to close the abnormally leaking blood vessels, to reset the retinal detachment of the macular area, and to prevent atrophy and degeneration^[9], we preferred retinal laser photocoagulation in the right eye, and anti-VEGF combined with laser in the left eye. VEGF can cause breakdown of the blood-retinal barrier, which induces

extracellular fluid accumulation in the macular region, anti-VEGF agents have been demonstrated to be effective in reducing vascular permeability and resolution of subretinal and intraretinal fluid in multiple ophthalmic pathologies. Not only that, if there is too much subretinal fluid, it will affect the thermal effect of the laser on the retina, creating an ineffective laser treatment. On the one hand, anti-VEGF treatment can reduce the subretinal fluid, on the other hand, it can make the subsequent laser treatment more effective, increase the adhesion between the retina and the deeper tissues, reduce the vascular leakage, and restore the anatomical structure of the retina. We then observed the patient for 6mo and the fundus condition improved dramatically. Consistent with those reported in the literature, if the patient has a large amount of subretinal fluid accumulation or a very large volume of CCH, anti-VEGF drugs can reduce vascular permeability, which can contribute to the rapid resolution of subretinal fluid and improve visual function^[10] and also reduce the risk of complications associated with other subsequent treatments in patients with CCH, and the overall prognosis is also better^[11].

In conclusion, intravitreal injection of anti-VEGF drugs combined with laser photocoagulation is a safe and effective treatment for choroidal hemangioma. Correct diagnosis and differential diagnosis are particularly important when relevant patients are seen in clinical practice, choosing the right approach to treat CCH earlier can also lead to better long-term outcomes.

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