Recalcitrant Circumscribed Choroidal Hemangioma Managed with Combination Therapy

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OBJECTIVE
To describe clinical course, imaging features and combined management of a recalcitrant circumscribed choroidal hemangioma.

PURPOSE
Circumscribed choroidal hemangiomas (CCH) are rare, solitary vascular hamartomas that have a predilection for the peripapillary area. Treatment modalities described have historically included laser photocoagulation, cryotherapy, cataract surgery, photodynamic therapy (PDT) and even intravitreal anti-VEGF. This report describes the clinical course of an aggressive CCH and its synergistic management with combination therapy.

METHODS
Retrospective, longitudinal chart review of a 38 year-old male with macular edema secondary to CCH. The patient was managed with a combination of half fluence verteporfin PDT (6 mg/m²/699 nm diode laser; 300 mW/cm²; 83 s), standard fluence verteporfin PDT (6 mg/m²/699 nm diode laser; 600 mW/cm²; 83 s) and intravitreal bevacizumab (IVB). The patient underwent periodic evaluations with ophthalmoscopy, spectral domain optical coherence tomography (SD-OCT), ultrasoundography, and angiography over the 3 year follow-up interval.

RESULTS
A 38 year-old otherwise healthy male with a chief complaint of blurry vision and metamorphopsia in his right eye presented with visual acuity of 20/50, macular edema and a large juxtapapillary choroidal lesion. A CCH was diagnosed (Figure 1 and Figure 2) and the patient successfully received half fluence verteporfin PDT. Although this resulted in resolution of the subretinal fluid, recurrence of macular edema was noted clinically after 6 months and the patient received three consecutive injections of IVB with good response and 20/20 vision (Figure 3). There was a 12 month interval before the patient experienced another recurrence of subretinal fluid and despite two consecutive IVB treatments, the vision decreased to 20/70 in the presence of angiographic and clinical CCH activity (Figure 4). The patient received standard fluence PDT with verteporfin and same-day intravitreal bevacizumab, with significant improvement in subretinal fluid (Figure 5) and visual acuity of 20/40 at the last follow-up.

CONCLUSION
Circumscribed choroidal hemangiomas are benign choroidal tumors that may develop vascular congestion over time and cause visually significant serous macular detachments. PDT has been shown to induce atrophy of these tumors and is becoming the treatment of choice, but it may not provide complete effect in the case of large tumors with thick overlying retinal detachments due to the limited ability to penetrate through the exudative fluid. Additionally, juxtapapillary tumors may not always be amenable to PDT or other laser treatment modality due to the risk of damage to the optic nerve.

It has been shown that intravitreal bevacizumab can improve visual acuity in cases of macular edema secondary to CCH. This case report demonstrates that when managing cases of refractory CCH, bevacizumab treatment may be used in conjunction with verteporfin PDT to decrease vascular hyperpermeability and holds promise in improving macular edema and final visual outcomes.

ACKNOWLEDGMENTS
The author has no financial conflicts of interest to declare.

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REFERENCES

Figure 1
B-scan ultrasonography reveals choroidal lesion superonasal to the optic nerve.
Base: 9.34 mm x 9.19 mm. Height: 2.29 mm. The lesion displays high-reflectivity on A-scan.

Figure 2
Color fundus photography of the right eye shows large, homogeneously appearing reddish-orange juxtapapillary CCH (left) that exhibits late diffuse hyperfluorescence with fine border of superior and nasal hypofluorescence on fluorescein angiography (right).

Figure 3
SD—OCT showing initially large serous macular detachment (left) with dramatic improvement of subretinal fluid and foveal re-attachment after treatment (right). Note bluer of residual subretinal fluid nasally (yellow arrow).

Figure 4
Vascularized CCH with cuff of subretinal fluid (left) that shows large hyperfluorescent inner tumor vascular channels both on the fluorescein (middle) and indocyanine green (right) angiographies. Note extensive macular involvement secondary to fluid leakage from the tumor (yellow arrow).

Figure 5
SD-OCT after 12 months showing recurring serous macular detachment (left) that decreases significantly after combined treatment (right).
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Circumscribed choroidal hemangiomas (CCH) are rare, solitary vascular hamartomas that have a predilection for the peripapillary area. Treatment modalities described have historically included laser photocoagulation, radiotherapy, cryotherapy, thermotherapy, and more recently, photodynamic therapy (PDT) and even intravitreal anti-VEGF. This report describes the clinical course of an aggressive CCH and its synergistic management with combination therapy.
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Retrospective, longitudinal chart review of a 38 year-old male with macular edema secondary to CCH. The patient was managed with a combination of half fluence verteporfin PDT (6 mg/m²; 689 nm diode laser; 300 mW/cm²; 83 s), standard fluence verteporfin PDT (6 mg/m²; 689 nm diode laser; 600 mW/cm²; 83 s) and intravitreal injections of 1.25 mg/0.05 mL bevacizumab (IVB). The patient underwent periodic evaluations with ophthalmoscopy, spectral domain optical coherence tomography (SD-OCT), ultrasonography, and angiography over the 3 year follow-up interval.
RESULTS

A 38 year-old otherwise healthy male with a chief complaint of blurry vision and metamorphopsia in his right eye presented with visual acuity of 20/50, macular edema and a large juxtapapillary choroidal lesion. A CCH was diagnosed (Figure 1 and Figure 2) and the patient successfully received half fluence verteporfin PDT. Although this resulted in resolution of the subretinal fluid, recurrence of macular edema was noted clinically after 6 months and the patient received three consecutive injections of IVB with good response and 20/20 vision (Figure 3). There was a 12 month interval before the patient experienced another recurrence of subretinal fluid and despite two consecutive IVB treatments, the vision decreased to 20/70 in the presence of angiographic and clinical CCH activity (Figure 4). The patient received standard fluence PDT with verteporfin and same-day intravitreal bevacizumab, with significant improvement in subretinal fluid (Figure 5) and visual acuity of 20/40 at the last follow-up.
CONCLUSIONS

Circumscribed choroidal hemangiomas are benign choroidal tumors that may develop vascular congestion over time and cause visually significant serous macular detachments. PDT has been shown to induce atrophy of these tumors and is becoming the treatment of choice, but it may not provide complete effect in the case of large tumors with thick overlying retinal detachments due to the limited ability to penetrate through the exudative fluid. Additionally, juxtapapillary tumors may not always be amenable to PDT or other laser treatment modality due to the risk of damage to the optic nerve. It has been shown that intravitreal bevacizumab can improve visual acuity in cases of macular edema secondary to CCH. This case report demonstrates that when managing cases of refractory CCH, bevacizumab treatment may be used in conjunction with verteporfin PDT to decrease vascular hyperpermeability and holds promise in improving macular edema and final visual outcomes.
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**FIGURES**

**Figure 1**
- B-scan ultrasonography reveals choroidal lesion superonasal to the optic nerve.
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- The lesion displays high-reflectivity on A-scan.

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- Color fundus photography of the right eye shows large, homogenously appearing reddish-orange juxtapapillary CCH (left) that exhibits late diffuse hyperfluorescence with fine border of superior and nasal hypofluorescence on fluorescein angiography (right).

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- SD-OCT showing initially large serous macular detachment (left) with dramatic improvement of subretinal fluid and foveal re-attachment after treatment (right). Note sliver of residual subretinal fluid nasally (yellow arrow).

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- Vascularized CCH with cuff of subretinal fluid (left) that shows large hyperfluorescent inner tumor vascular channels both on the fluorescein (middle) and indocyanine green (right) angiographies. Note extensive macular involvement secondary to fluid leakage from the tumor (yellow arrow).

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