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REPAIR OF COMBINED TRACTION-RHEGMATOGENOUS RETINAL DETACHMENT AFTER CRYOABLATION OF A RETINAL CAPILLARY HEMANGIOBLASTOMA

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Purpose: To describe a case of surgical repair of a total, combined traction-rhegmatogenous, retinal detachment with proliferative vitreoretinopathy after cryoablation of a retinal capillary hemangioblastoma.

Methods: A case of a 47-year-old man presenting with a solitary, superotemporal retinal capillary hemangioblastoma in the right eye with serous retinal detachment and subfoveal fluid and exudates is reported. The hemangioblastoma was treated with cryoablation, but despite regression of the lesion, the patient developed a total combined traction-rhegmatogenous retinal detachment 6 weeks later. Vitrectomy, endolaser photocoagulation to tears adjacent to the original hemangioblastoma lesion, and silicone oil exchange was performed to repair the detachment.

Results: Eighteen months after initial repair, the patient had silicone oil removal and cataract extraction with lens implantation. Final visual acuity improved from counting fingers to 20/50 with total retinal reattachment and regression of the retinal capillary hemangioblastoma.

Conclusion: Although uncommon, combined traction-rhegmatogenous retinal detachment can occur after cryoablation of a retinal capillary hemangioblastoma.

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Retinal capillary hemangioblastomas (RCHs) are benign vascular tumors that can diminish visual acuity by exuding subretinal fluid or by causing tractional forces that can lead to retinal detachment (RD) or other anatomical distortion of the retina.[1] Cryoablation (also called cryotherapy) of RCH is a standard treatment option for lesions of appropriate size and location. Exudative RD is a relatively uncommon complication of cryoablation.[2] We report a striking case of a total rhegmatogenous and tractional RD after cryotherapy of a sporadic, isolated RCH and the results of subsequent, successful, surgical repair.

Case Report

A 47-year-old man presented with progressively worsening visual acuity in the right eye and was noted to have a 3-mm RCH in the superotemporal midzone with associated serous RD, macular exudates, and subfoveal fluid (Figure 1A). The subfoveal fluid was well-demonstrated on optical coherence tomography (Figure 2). The patient's visual acuity in the right eye was 20/70. He underwent cryoablation with a double freeze–thaw technique. Two days later, visual acuity improved to 20/25 (Figure 1B). A brain and spine magnetic resonance imaging study was performed and was negative for any abnormality associated with von Hippel–Lindau (VHL) disease. Renal cell carcinoma and

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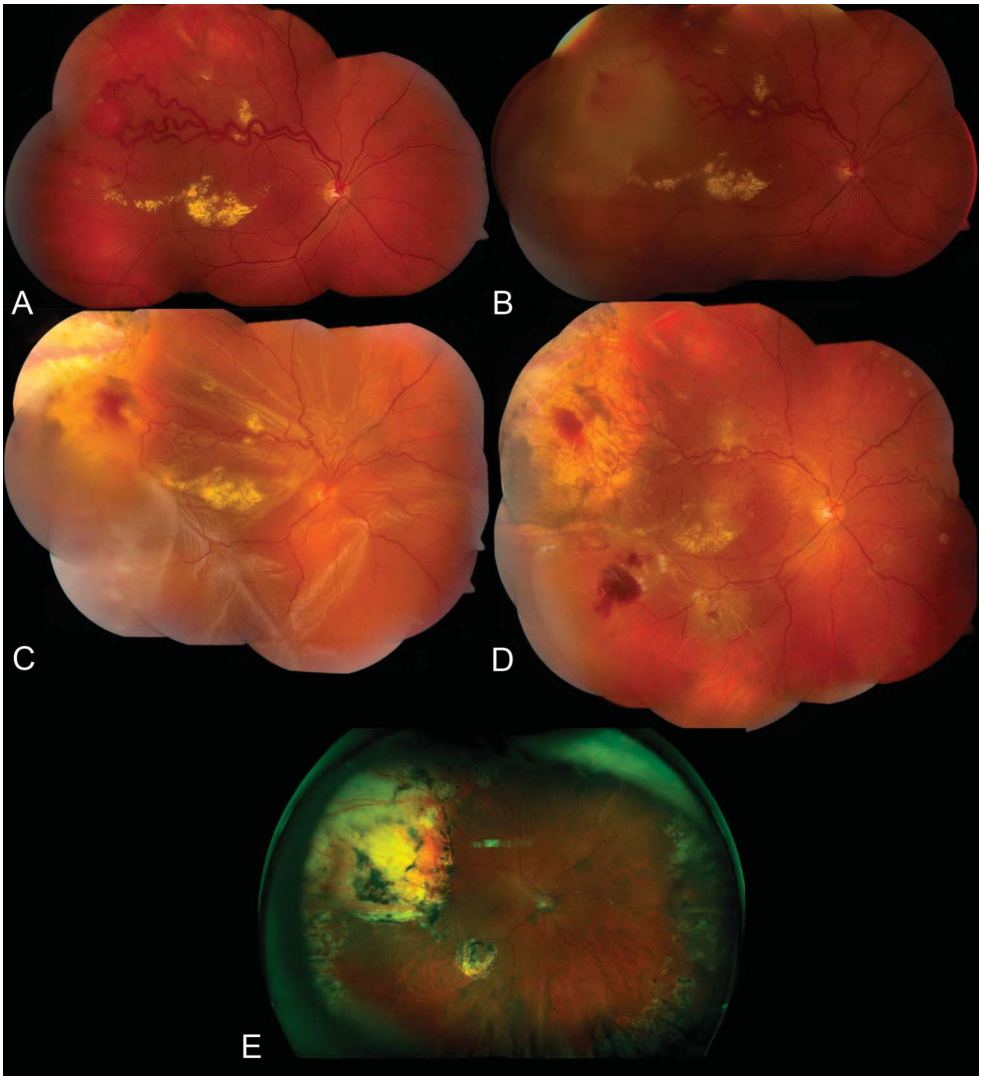

Fig. 1. Color fundus photography of the right eye of the patient. **A.** Before any treatment. **B.** Two days after cryoablation of the RCH. **C.** Six weeks after treatment, the patient developed a total combined traction-rhegmatogenous RD with proliferative vitreoretinopathy. **D.** Two weeks after RD surgical repair, which included endolaser photocoagulation and silicone oil injection. **E.** Two years after surgical repair, after having had silicone oil removal and cataract extraction with lens implantation.

pheochromocytoma workup, including a urine metanephrine study, were negative. Subsequent genetic testing for VHL was negative.

Despite initial improvement and continued regression of the hemangioblastoma, the patient developed a total combined traction-rhegmatogenous RD six weeks later with proliferative vitreoretinopathy (Figure 1C) and visual acuity of count fingers. Vitrectomy with membrane peeling, retinotomy, drainage of the subretinal fluid, endolaser photocoagulation, and silicone oil injection was performed. The hemangioma regressed, the macula flattened, and the dilated feeder vessels resolved 2 weeks later (Figure 1D). Eighteen months later, the patient had a combined silicone oil removal and cataract extraction with lens implantation; visual acuity improved to 20/50. Peripheral retinal scarring at the site of cryotherapy, along with a flat retina, was noted 28 months after the initial treatment of the hemangioblastoma (Figure 1E).

Discussion

Retinal capillary hemangioblastomas are benign vascular hamartomas that generally have clinical onset

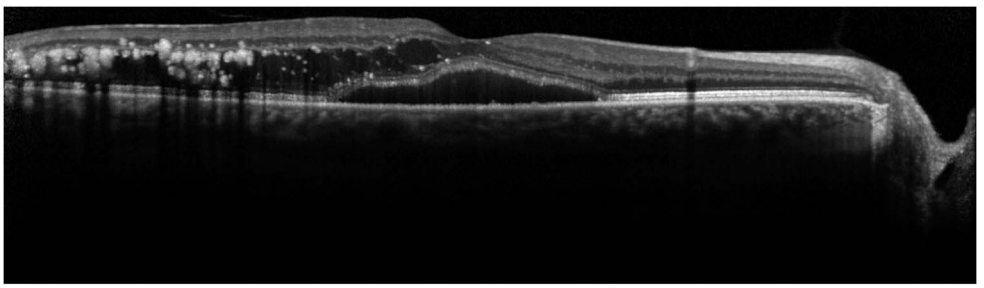

Fig. 2. Enhanced depth imaging optical coherence topography of the right eye before any treatment of the RCH; a horizontal line scan centered at the fovea demonstrates subfoveal fluid and macular exudates contributing to decreased visual acuity.

within the first 2 decades of life.[1,3] von Hippel–Lindau is a rare (1/36,000 live births), autosomal dominant disease that predisposes to hemangioblastomas of the retina and central nervous system, pancreatic carcinoma, pheochromocytomas, and renal cysts.[4,5] Bilateral or multiple RCH are strongly associated with VHL, while solitary RCH may be sporadic or associated with VHL.[3] Retinal capillary hemangioblastoma is the earliest and most frequent manifestation of VHL.[6] Therefore, ophthalmologists must coordinate appropriate testing for systemic features of VHL when RCH is initially detected. Our patient had prompt neuroimaging and laboratory studies that were unremarkable, and genetic testing was negative for VHL. Genetic testing is important to guide appropriate surveillance. The Cambridge protocol indicates that patients with genetically confirmed disease require annual retinal examination and a brain magnetic resonance imaging every 3 years until the age of 50, and every 5 years thereafter.[7–9] Appropriate discussion regarding the autosomal dominant inheritance pattern of VHL should be given to patients with the genetically confirmed disease.

Our patient demonstrated the two major visually consequential features of RCH. The first is fluid exudation, which can lead to macular exudates and serous RD. The second is glial proliferation, which can contribute to the development of a combined traction-rhegmatogenous RD.[10] These threats to visual acuity, punctuated by their proximity to the fovea, were clear indications for treatment in our patient. Numerous treatment options have been described, including monotherapy or combination therapy of cryoablation, focal photocoagulation (FP), transpupillary thermotherapy, photodynamic therapy, radiotherapy, anti–vascular endothelial growth factor (anti-VEGF) intravitreal injection, triamcinolone intravitreal injection, or vitreoretinal surgery with lesion excision or feeder vessel ligation.[10–17] Choice of treatment modality is generally guided by tumor characteristics, such as location, size, extent of complications, and surgeon preference. Focal photocoagulation and cryotherapy remain mainstays of treatment; they have been shown to be equally efficacious to one another and have a long-track record of success. Focal photocoagulation has been shown to be particularly successful when the RCH is peripheral and 1.5 mm or smaller in size, whereas cryotherapy is usually considered for larger lesions, up to about 4.5 mm.[10,11,18]

We chose cryotherapy because of our previous experience with good results in lesions up to 3 mm. Cryotherapy is a particularly useful when there is significant subretinal fluid that may limit laser energy uptake as in this case. We used a conventional double freeze–thaw cycle to the hemangioma itself, but also applied one cycle anterior to the lesion. *Ablatio fugax* is the worsening of subretinal fluid and transudative RD following ablation. It is more often associated with FP when high, concentrated laser energy is used on large lesions and believed to be less common after cryotherapy.[13,19] Although there are reports of *ablatio fugax* after cryotherapy of other vasoproliferative disorders, such as Coats disease and retinopathy of prematurity, our report of this complication after cryotherapy of an RCH is a relatively unique contribution to the literature.[2,20,21] It is believed that the fluid exudation, perifoveal lipid exudation, and cystoid macular edema associated with *ablatio fugax* can be prevented by using local or systemic corticosteroids to inhibit the inflammatory response that occurs after ablative destruction of retinal tissue.[22,23] Use of corticosteroids may have helped prevent these complications from developing in this case and may be a particularly helpful adjunct when cryoablation is heavily applied.

Overall, the total RD requiring vitrectomy seen in our patient is rare, especially after cryotherapy. Visual outcome after treatment of RCH is variable, depending on size and location of the tumor and the resultant degree of exudation and traction. In one large series, 40% of RCH treated with FP, cryotherapy, or diathermy had a visual prognosis $<20/200$.[24] Our patient fared better despite the severity of the RD complication.

In conclusion, although cryotherapy is usually effective in halting progression of RCH, patients must be carefully monitored for increased retinal exudation and the rare possibility of total RD after treatment. Patients must be carefully counseled on the vision-threatening nature of the condition, and the potential risks and sequelae of treatment.

Key words: capillary hemangioblastoma, capillary hemangioma, retinal cryoablation, retinal cryotherapy, retinal detachment, retinal hemangioblastoma, von Hippel, von Hippel–Lindau.

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