# An Unusual Case of Retinal Hemangioma Presenting with Retinal Hole and Tear

## Retinal Yırtık ve Delikle Seyreden Sıradışı Bir Retinal Hemanjiom Vakası

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*Case Report* *Olgu Sunumu*

**ABSTRACT**

Retinal capillary hemangioma (RCH) is commonly observed in von Hippel-Lindau (VHL) disease. Many complications have been reported due to RCHs; exudative or tractional retinal detachment, vitreous hemorrhage, cataract, iris neovascularization are among the most commonly reported. We report an unusual case of unilateral RCH as a part of VHL disease presenting with a concomitant retinal hole and a retinal tear.

**Key Words:** Retinal capillary hemangioma, von Hippel-Lindau disease, retinal tear, retinal hole.

**ÖZ**

Retinal kapiller hemanjiom (RKH), Von-Hippel Lindau (VHL) hastalığında sık görülen bir bulgudur. Günümüze kadar RKH'lara bağlı çok sayıda komplikasyon bildirilmiştir; bunlar arasında, eksudatif ya da traksiyonel retina dekolmanları, vitre hemorajisi, katarakt,iris neovaskülarizasyonu en sık söz edilenlerdir. Çalışmamızda , VHL hastalığı ile birlikte giden, beraberinde retinal delik ve yırtıkla seyreden tek taraflı RKH olgusunu sunduk.

**Anahtar Kelimeler:** Retinal kapiller hemanjiom, von Hippel-Lindau hastalığı, retinal yırtık, retinal delik.

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## INTRODUCTION

RCHs are the benign vascular tumors of the retina or the optic nerve head, usually occuring sporadically or as a part of VHL syndrome

VHL is caused by a germline mutation in the VHL gene that has been mapped to chromosome 3p25.[1] The incidence is reported to be about one in 36000 live births and most patients present between 10 to 40 years of age. Twenty percent of patients appear with no family history accounting for the de novo mutations. The cardinal features of the syndrome are RCH, central nervous system hemangioblastomas, renal cell carcinomas, phechromocytomas and pancreatic, renal and epididymal cysts. RCH is the most common and the earliest presentation of the syndrome.[2,3]

The vision loss results from exudation of fluid into the macula, glial proliferation or vitreous hemorrhage. The treatment options described for RCH are photocoagulation, cryotherapy, transscleral diathermy, photodynamic therapy and transpupillary thermotherapy.[4]

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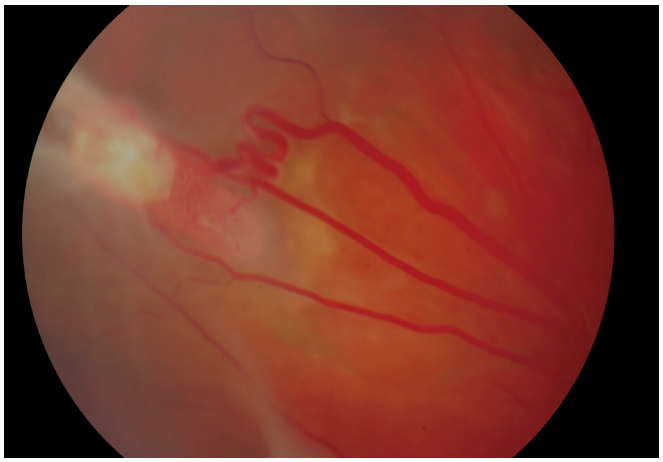

**Figure 1:** Fundus photo before treatment.

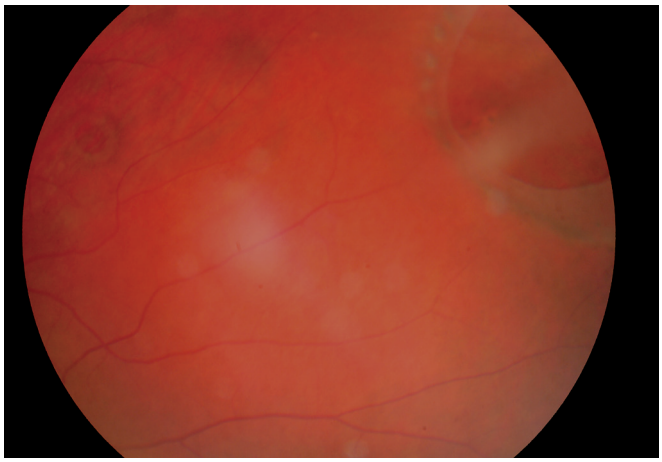

**Figure 2:** Fundus photo of the retinal tear and the atrophic retinal hole.

## CASE REPORT

A 37 year old woman referred from the neurosurgery clinic where 20 days ago she has been operated for a cerebellar hemangioblastoma with the diagnosis of Von Hippel Lindau disease. She had no ocular complaints at presentation. Her past medical history was insignificant apart from the cerbellar hemangioblastoma which has been operated recently. Her family history was significant with three elder sisters having had neurosurgical operations, one with an accompanying renal tumor.

On examination she had 20/20 vision on both eyes with normal anterior segment. The intraocular pressure was within normal limits bilaterally. On fundoscopy, right eye was normal, left eye showed an endophytic RCH of 2 disc diameter located superonasally about 5 disc diameters away from the papilla (Figure 1). The large feeding and draining vessels were observed near the lesion with a local tractional retinal detachment.

An atrophic retinal hole of 500 micron diameter was located on the superotemporal arcade neighboring a retinal tear at the superotemporal equatorial region(Figure 2). Fundus fluorescein angiography of the left eye revealed early hyperfluorescence with late leakage (Figure 3). Focal treatment was also applied along the borders of the retinal hole and the tear at the first laser session.

Focal laser treatment over the lesion and around the tractional retinal detachment was applied 5 times through the 6 months follow-up at the end of which sub-total involution of the tumor was achieved and the subretinal fluid was mostly absorbed (Figure 4).

The visual acuity was still 20/20 on both eyes. Laser parameters for the lesion was 1000 mW intensity, 200 micron spot size and 0.2 second duration. The parameters for the peripheral hole and tear were 350 mW, 300 microns and 0.1 seconds. During the course of the follow-up, the patient had an abdominal MRI which revealed bilateral renal lesions strongly suggestive of renal cell carcinoma and she was referred to another center for the treatment of this expected finding.

## DISCUSSION

VHL disease is an autosomal dominant disorder hence for an affected individual, there is a 50 percent risk to transmit the disease to his/her offspring. Our patient had three elder sisters previously diagnosed with cerebellar tumors and one with a renal tumor, the records of these could not be obtained. Given the strong family history and 3 visceral organ tumors our patient was diagnosed with VHL disease and no further genetic testing was conducted.

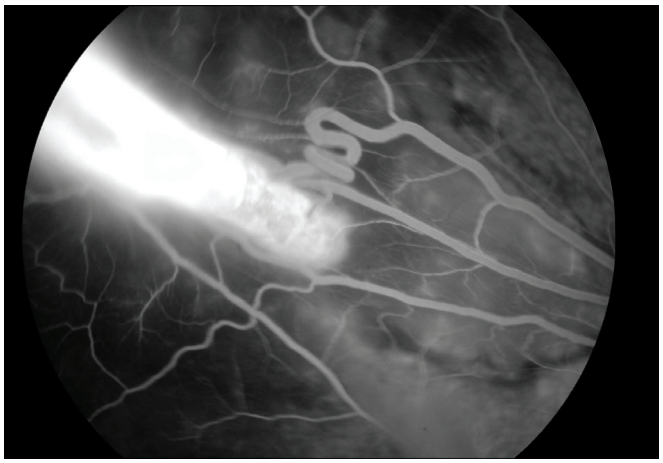

**Figure 3:** FFA image before treatment.

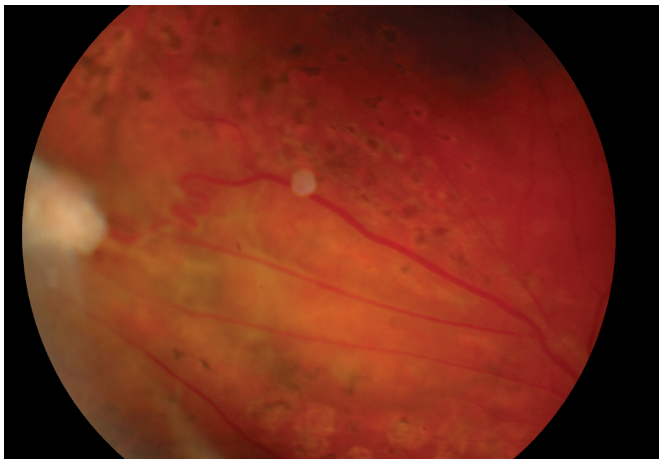

**Figure 4:** Fundus photo at the 6$^{th}$ month of treatment.

The RCH in VHL disease is a slow growing tumor which is a cellular compound of stromal and endothelial cells, pericytes and mast cells. Vision loss occurs secondary to complications of the tumor, mainly exudation and glial proliferation. Our case presented with a small area of tractional detachment secondary to the glial proliferation over the lesion. Various complications and accompanying findings have been reported with RCHs; which exudative retinal detachments, tractional retinal detachments, vitreous hemorrhages, iris neovascularization, macular hole and cataract are among the most common. To our knowledge, this is the first reported case of RCH presenting with a retinal hole and a tear.

Different treatment modalities have been described. Photocoagulation and cryotherapy are main treatment options and multiple treatments may be necessary to fully obliterate the lesion. In large tumors, extensive photocoagulation or cryotherapy may induce retinal exudation. Pre and post op therapy with corticosteroids may reduce this unwanted complication.

In our case no negative effect was observed with the application of laser photocoagulation of this large retinal tumor. There are limited number of studies on the possible use of systemic or intravitreal anti-VEGF therapy for RCH. So far, they have found that anti-VEGF therapy reduces the amount of exudation and may improve vision but does not change the size of the lesion.[5] Photodynamic therapy with vertoporfirin has been reported to be successful by many authors.[6] Plaque radiotherapy, transpupillary thermotherapy and vitreoretinal surgery have also been used in selected cases.

However none of these have been studied in a randomized controlled study. In our case, direct and peripheral lesional photocoagulation resulted in continuous improvement in terms of lesion size and subretinal fluid absorbtion and at the end of the 6$^{th}$ session the tumor was regressed and absorbtion of a major part of subretinal fluid was achieved.

Our case has presented with a concomitant retinal hole and a retinal tear both of which were located on the temporal periphery where no tractional force was observed as the tumor itself was located on the nasal side. No exudation under the hole or the tear has been noted. Whether these findings were a complication of the tumor or coincidental is a subject that needs to be further evaluated through more future reported cases.

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