Transpupillary Thermotherapy for Retinal Capillary Hemangiomas in Von Hippel-Lindau Disease

Von Hippel-Lindau Hastalığında Retinal Kapiller Hemanjiyomların Transpupiller Termoterapi ile Tedavisi*

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Case Report *Olgu Sunumu*

ABSTRACT

A 22-year-old woman presented with a 5-day history of reduced vision in her left eye. Fundus examination revealed multiple different sized retinal capillary hemangiomas (RCH) in her left eye. Serous retinal elevation and intraretinal edema surrounding the RCHs in the superior retinal periphery extended to the macula. The moderate and large sized RCHs were treated with transpupillary thermotherapy (TTT) with 810 nm diode laser. The macular exudation resolved rapidly. In the follow-up, additional TTT was applied to completely obliterate the large sized RCHs, which were additionally circumscribed with 532 nm laser photocoagulation. All RCHs scarred completely. The visual acuity increased to 20/20 at 6 months, and remained stable throughout the 30-month follow-up period. TTT may have a therapeutic role in the management of selected RCH cases, particularly in cases presenting with multiple and large sized extrapapillary tumors.

Key Words: Capillary hemangioma, retina, transpupillary thermotherapy, von hippel-lindau disease.

ÖZ

Yirmi iki yaşındaki kadın hasta sol gözünde 5 gündür olan görme kaybı ile başvurdu. Fundus muayenesinde, sol gözünde birden çok, farklı boyutlarda retinal kapiller hemanjiyom (RKH) görüldü. Süperior periferik retinadaki seröz elevasyon ve intraretinal ödem makülaya kadar uzanmaktaydı. Orta ve büyük boy RKH'ler 810 nm diyot laser ile transpupiller termoterapi (TTT) ile tedavi edildi. Makuladaki eksüdasyon hızlıca düzeldi. Takiplerde, büyük boy RKH'lere ek TTT uygulanarak, tam kapanmaları sağlandı. Ayrıca büyük boy RKH'ler 532 nm laser ile çevrelendi. Tüm hemanjiyomların tamamen skarlaştığı görüldü. Görme düzeyi 6. ayda 20/20'ye ulaştı ve 30 aylık takip boyunca sabit kaldı. TTT seçilmiş RKH olgularında, özellikle çok sayıda ve büyük hemanjiyomların tedavisinde yararlı bir yöntem olabilir.

Anahtar Kelimeler: Kapiller Hemanjiyom, retina, transpupiller termoterapi, von hippel-lindau hastalığı.

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INTRODUCTION

Von Hippel-Lindau (VHL) disease is an autosomal dominant life threatening phacomatosis characterized by various tumors in multiple organs. The most common and earliest manifestation is retinal capillary hemangioma (RCH), which usually presents with loss of vision during the 2-3rd decade.[1]

The treatment of RCH can be challenging, especially in the presence of bilateral and multiple tumors and with the continuous development of new tumors.[2]

Transpupillary thermotherapy (TTT) is a well-established means of treatment for intraocular tumors. However, there is limited experience with TTT in the treatment of RCH, and therefore its role in treatment of RCHs is still mostly unknown.[2-6]

The aim of this report is to describe the use of TTT with 810 nm diode laser in the treatment of a challenging patient with VHL disease, who presented with multiple different sized, rapidly progressive RCHs with macular exudation severely affecting central vision.

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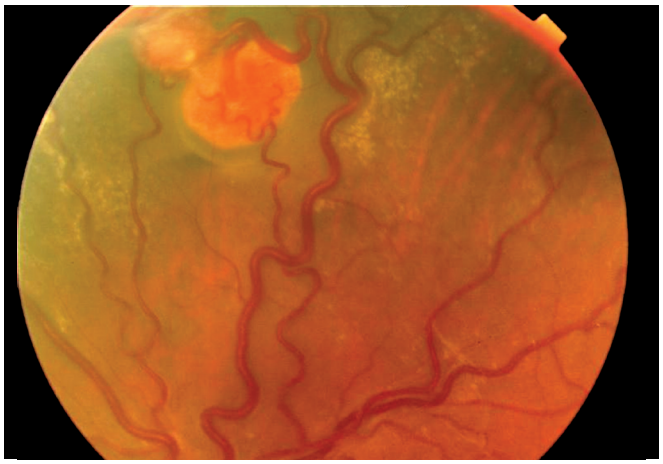

Figure 1: Superior peripheral retina of the left eye at presentation. A large sized RCH with surrounding intraretinal edema and serous elevation is observed. The feeder vessels appear dilated and tortuous.

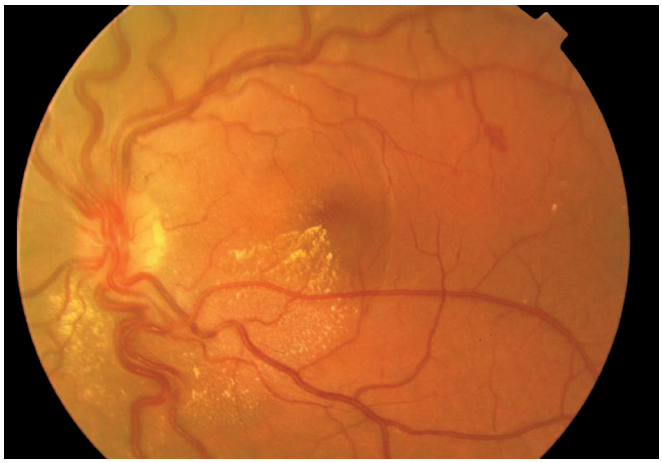

Figure 2: The macula of the left eye at presentation. Intraretinal edema with hard exudates and serous elevation are observed. Dilated and tortuous feeder vessels are seen.

CASE REPORT

A 22-year-old woman presented with progressive loss of visual acuity (VA) over 5 days in her left eye. The best-corrected VA was 20/20 and 20/200 in her right and left eyes, respectively. Fundus examination revealed one large sized (greater than 3.0 mm) RCH in the right and three in the left eye (Figure 1). Additionally, there were two moderate sized (1.5-3.0 mm) RCHs in the left eye, and various small (smaller than 1.5 mm) RCHs in both eyes. All of the RCHs were located outside the temporal arcuates in the left eye. However, serous retinal elevation surrounding the RCHs in the superior periphery extended inferiorly leading to intraretinal edema and serous elevation at the macula (Figure 2). In fluorescein angiography, rapid filling in the arterial phase and leakage in the late phase were documented.

The moderate and large sized RCHs in both eyes were treated with TTT with 810 nm diode laser (IRIS Oculight SLx, IRIS Medical Inc, Mountain View, CA). The laser parameters were adjusted to obtain moderate whitening of each lesion beginning with 800 mW power, 60 seconds pulse duration for 3000 μm spot size (10 W/cm^2). The other small sized RCHs were treated with 532 nm laser photocoagulation. Although a slight increase in retinal exudation in the left eye was observed in the first 2 days, the exudation resolved rapidly in the following 2 weeks, and the macula was totally dry at 1 month (Figure 3). The VA increased to 20/60 at 2 months and to 20/20 at 6 months.

At 1 month, additional TTT was applied to completely obliterate the large sized RCHs, which were also circumscribed with 532 nm laser photocoagulation. At 6 months, all RCHs had totally scarred without any exudation (Figure 4). Further laser photocoagulation was applied only to new developing RCHs. Although fibrous proliferation developed from scarred large sized RCHs in 6 months, retinal detachment did not occur. At 30 months, the VA in both eyes was preserved. Systemic investigation revealed hemangiomas in the cerebellum and medulla oblongata, syringomyelia, renal cell carcinoma in the right kidney, multiple renal and pancreatic cysts. No sign of disease could be disclosed among the family members.

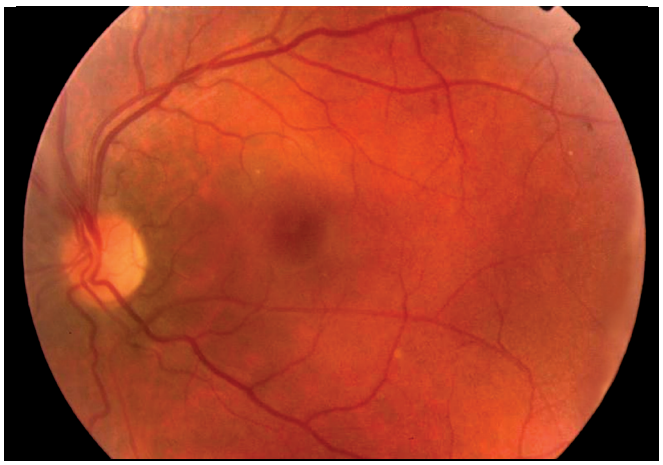

Figure 3: The macula of the left eye at 1 month after TTT. The edema, exudates and serous elevation have resolved. The dilatation and tortuosity of feeder vessels have regressed.

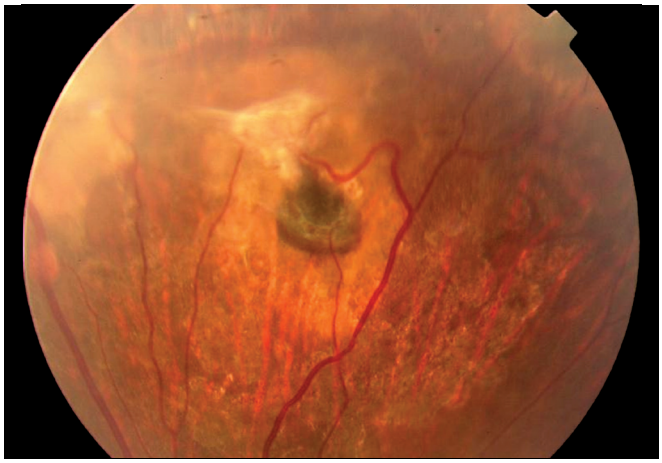

Figure 4: The RCH in figure 1 at 6 months. The tumor scarred totally, dilatation and tortuosity of vessels have regressed. The 532 nm laser photocoagulation provided a firmly attached retinal zone limiting tractional effects.

DISCUSSION

The decision and modality to treat a RCH depend on the size and location of the tumor, the presence and extent of associated findings such as exudation or traction. Most universal methods of management are observation, laser photocoagulation, cryotherapy, plaque radiotherapy, and vitreoretinal surgery.[2]

RCHs can be classified as small (smaller than 1.5 mm), moderate-sized (1.5-3 mm) and large-sized (greater than 3 mm) based on the diameter of the tumor.[5] Thermal laser photocoagulation is most successful for RCHs with a diameter less than 1.5 mm^2. In this case, the moderate and large sized RCHs were not amenable to laser photocoagulation due to serous retinal elevation and their large size. In addition, compared to TTT laser photocoagulation induces more damage to the sensory retina and probably more retinal traction.[7]

An alternative therapeutic approach for this case could be cryotherapy. However, cryotherapy is a destructive approach causing extensive damage to surrounding tissues, inducing extensive scarring and proliferative vitreoretinopathy.[8] Cryotherapy has to be performed in operation room settings. Furthermore, postoperative lid edema, chemosis, and elevated intraocular pressure are frequent sources of discomfort for the patient. Therefore, cryotherapy was reserved as a second line approach in this case.

TTT was utilized in this case for various reasons. The adjustable spot size, power settings and laser duration provides the opportunity to treat different sized RCHs with a single spot with safe margins adjusted slightly larger than the tumor. In addition, it is possible to carry out the treatment in office settings, with practically no discomfort to the patient. This also provides the opportunity for additional treatments without difficulty.

In this case, the resolution of the retinal exudates began shortly after the first application of TTT. With close follow-up, it was possible to accelerate the scarring of large tumors with additional TTT. The large sized RCHs treated with TTT were also circumscribed with 532 nm laser photocoagulation after two months. This provided a firmly attached retinal zone limiting tractional effects of proliferating fibrous bands which could result in tractional retinal detachment.

Previously, Parmar et al., utilized TTT for juxtapapillary RCHs in three cases. They observed complete fibrosis of the tumor in only one case; however it was associated with optic atrophy.[3] Similarly, Garcia-Arumi et al. utilized TTT for two cases of juxtapapillary RCH. They did not note any complications, but neither any efficacy.[6]

Mochizuki et al. reported successful results after TTT for two cases of extrapapillary RCHs. However, they reported a minor complication; bleeding on tumor surface after application of high energy (10 W/cm^2 for 6 minutes) in one eye.[4] Conversely, we utilized the same energy level (10 W/cm^2) for a shorter duration (60 seconds), and did not encounter any complication.

These favorable results of TTT in the treatment of extrapapillary RCHs are encouraging, and warrant gathering of more RCH cases and evaluation of longer term results.

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