Available online at www.sciencedirect.com

ScienceDirect

Journal of the Chinese Medical Association 77 (2014) 52–56

www.jcma-online.com

Case Report

Persistent exudative retinal detachment after photodynamic therapy and intravitreal bevacizumab injection for multiple retinal capillary hemangiomas in a patient with von Hippel–Lindau disease

Fang-Yi Tsai [a], Ling-Ing Lau [a,b,c,*], Shih-Jen Chen [a,b], Fenq-Lih Lee [a,b], Shui-Mei Lee [a,b]

[a] *Department of Ophthalmology, Taipei Veterans General Hospital, Taipei, Taiwan, ROC*
[b] *Department of Ophthalmology, National Yang-Ming University School of Medicine, Taipei, Taiwan, ROC*
[c] *Institute of Clinical Medicine, National Yang-Ming University School of Medicine, Taipei, Taiwan, ROC*

Received September 22, 2011; accepted May 9, 2012

Abstract

Photodynamic therapy (PDT) has been used in treating peripheral retinal capillary hemangioma (RCH) with satisfactory results. We report a rare case of von Hippel-Lindau (VHL) disease with three large peripheral RCHs, treated with PDT and intravitreal bevacizumab injection (IVB), who developed persistent bullous exudative retinal detachment (RD) despite significant tumor regression. The patient is a sporadic case of VHL disease, with a *de novo* nonsense mutation in codon 161 with C → T transition at nucleotide position 694 of the *VHL* gene. Multiple RCHs were noted in both eyes. Four small RCHs were found in the left eye and were treated with laser photocoagulation. Three large RCHs in the peripheral retina of the right eye were complicated with cystoid macular edema and subretinal fluid accumulation. The RCHs were treated with PDT combined with IVB, and bullous exudative RD developed on the second day after treatment. Three months after PDT, the tumors had regressed significantly, but exudative RD persisted, despite multiple IVB and intravitreal triamcinolone acetonide injection (IVTA). External drainage with sclera buckling, IVB, and IVTA were performed, and the retina attached after surgical intervention. The application of PDT in the treatment of RCHs and its possible complications are discussed.

Keywords: exudative retinal detachment; photodynamic therapy; retinal capillary hemangioma; von Hippel–Lindau

1. Introduction

Retinal capillary hemangioma (RCH) is a benign vascular tumor which may occur sporadically or as a manifestation of von Hippel-Lindau (VHL) disease.[1] The visual prognosis is usually unfavorable, due to exudation from the tumor, causing intraretinal edema or exudative retinal detachment (RD). According to its location, RCH can be classified as a peripheral or a juxtapapillary type. Current treatment modalities for RCH include laser photocoagulation, cryotherapy, radiotherapy, traspupillary thermotherapy, and vitreoretinal surgery.[1,2] Laser photocoagulation and cryotherapy are the two major conventional therapies, and are effective as the sole method of treatment in controlling 74% and 72% of peripheral tumors, respectively.[2] However, larger tumors need multiple sessions of therapy, which may cause extensive exudative RD. In recent years, photodynamic therapy (PDT) has been used in treating large peripheral RCH with satisfactory results.[3] Most tumors regress with decreased exudation. We report a rare case of VHL disease, with three large peripheral RCHs treated with PDT and intravitreal bevacizumab injection (IVB), who developed persistent bullous exudative RD despite significant tumor regression.

* Corresponding author. Dr. Ling-Ing Lau, Department of Ophthalmology, Taipei Veterans General Hospital, 201, Section 2, Shih-Pai Road, Taipei 112, Taiwan, ROC.
E-mail address: leliew@vghtpe.gov.tw (L.-I. Lau).

1726-4901/$ - see front matter
http://dx.doi.org/10.1016/j.jcma.2013.09.001

2. Case report

A 23-year-old Taiwanese female had blurred vision in the right eye for 5 months. She had a past history of multiple intramedullary hemangiomas and had received two neurosurgeries to excise tumors at 14 years old and 22 years old. Reviewing the medical history of her family members, there was no known history of VHL disease or associated tumors. Genotyping of the *VHL* gene was performed after obtaining informed consent from the patient and her family members. Genomic DNA was extracted from peripheral venous blood samples. The three exons of the *VHL* gene were amplified by polymerase chain reaction (PCR) and sent to the National Yang-Ming University Genome Research Center for genotyping.

The presenting best-corrected visual acuity (BCVA) was 3/60 in the right eye and 6/5 in the left eye. There were no cells or flare in the anterior segment. Fundus examination revealed three large peripheral RCHs in the right eye, with prominent feeding arteries and tortuous dilated draining veins, complicated with subretinal fluid accumulation (Fig. 1A). Optical coherent tomography (OCT) showed marked cystoid macular edema and subretinal fluid accumulation at the macula (Fig. 2A). Fluorescein angiography of the tumors showed early hyperfluorescence with profound late leakage (Fig. 3). There were also four small peripheral RCHs in the left eye (Fig. 4A and B). The tumors in the left eye were treated with focal photocoagulation and regressed to gliotic nodules (Fig. 4C).

After obtaining informed consent, IVB 2.5 mg was administered to the right eye. Subretinal fluid was decreased

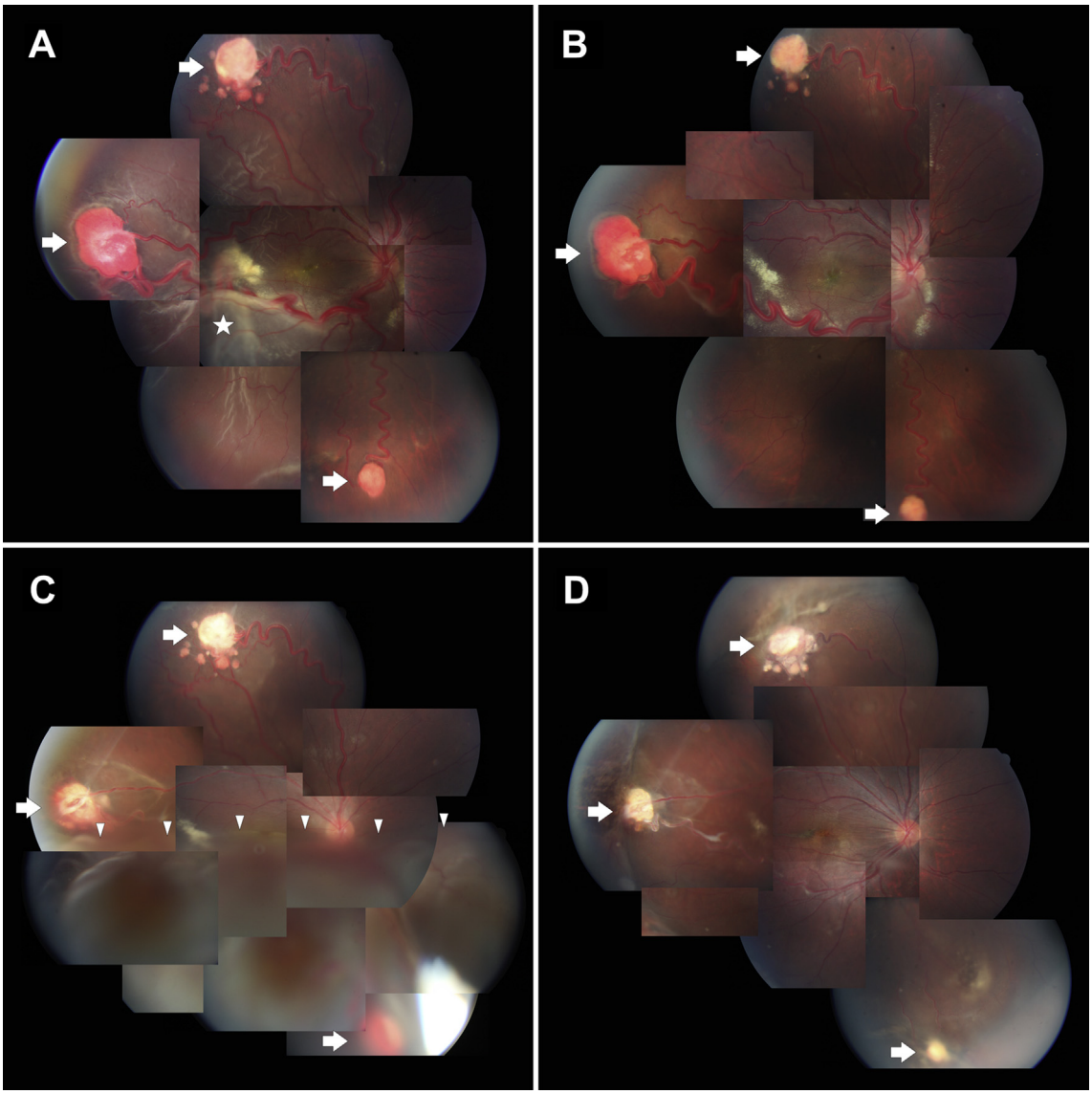

Fig. 1. (A) Three large retinal capillary hemangiomas in the peripheral retina in the right eye (arrows) with subretinal fluid accumulation (star) and lipid exudate at the macula; (B) decreased subretinal fluid and lipid exudate were noted at 1 week after 2.5 mg intravitreal bevacizumab injection; (C) 3 months after photodynamic therapy and intravitreal bevacizumab injection, the tumors showed significant regression (arrows). However, massive exudation with bullous retinal detachment persisted (below arrowheads) despite oral prednisolone and intravitreal injection of bevacizumab and triamcinolone acetonide; and (D) gliotic tumors (arrows) with resolved subretinal fluid at 3 months after surgical intervention.