

Case Report

MANAGEMENT OF PAPILLARY RETINAL CAPILLARY HEMANGIOMAS - CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Background: Retinal capillary hemangiomas (RCH) are rare, benign, vascular tumours characterized by capillary and glial cell proliferation. We report the management of a patient with a unilateral endophytic papillary RCH with exudations and serous retinal detachment.

Methods: Case report

Results: A 46-year old female presented with papillary RCH without association with von Hippel-Lindau syndrome. She was given two doses of intravitreal bevacizumab following which the serous detachment resolved. When the edema recurred again, she was given a dose of 4mg intravitreal triamcinolone acetate. The edema resolved and did not recur till the end of three months. She was given the option of photodynamic therapy, however, she did not consent for the procedure due to the high costs involved.

Conclusion: Intravitreal injections can be used as a temporary remedy for the treatment of macular edema caused by RCH. However, currently there is no ideal definitive treatment available for papillary RCH. Larger studies are necessary to establish the treatment for this condition.

Keywords: Avastin - Optic nerve head – Papillary - Photodynamic therapy (PDT) - Retinal capillary hemangiomas (RCH).

Introduction

Retinal capillary hemangiomas (RCH) are rare, benign, vascular tumours characterized by capillary and glial cell proliferation. These tumours can either be sporadic or associated with von Hippel-Lindau disease. Depending on their growth, they can be classified as endophytic, sessile and exophytic. They are mostly located in periphery or mid-periphery, and rarely near or on the optic disc. [1,2] Papillary RCH can lead to severe visual loss due to macular exudation and serous retinal detachment. Although peripheral RCH can be ablated

with laser photocoagulation, papillary RCH are difficult to treat. [3,4] Multiple therapeutic approaches including photodynamic therapy (PDT) and intravitreal anti-vascular endothelial growth factor (VEGF) injections have been reported. [1-5] However, the effectiveness of these treatments is limited. We report the management of a patient with a unilateral endophytic papillary RCH with exudations and serous retinal detachment.

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Case report

A 46-year old female presented with complaint of painless, progressive decrease in vision in left eye since last one month. She had no systemic complaints. Her best-corrected visual acuity (BCVA) was 20/20 in right eye and 20/200 in left eye. Ocular examination revealed round and normal reacting pupils with no relative afferent pupillary defect (RAPD); and parallel visual axes. Anterior segment examination in both the eyes was unremarkable. Posterior segment examination of right eye was normal; while left eye showed a well-defined yellowish-red lesion overlying the disc, completely obscuring its view and associated with serous macular detachment (Figure 1a). B scan showed an acoustical solid mass with smooth anterior border and variable medium reflectivity (Figure 1b). Fundus fluorescein angiography showed early hyperfluorescence, gradually

increasing intensity in mid-phase and gradually decreasing intensity in late phase (Figure 2). Optical coherence tomography showed hyperreflective mass overlying the disc (Figure 3a) with serous retinal detachment at the macula (Figure 3b). Findings were suggestive of a papillary retinal capillary hemangioma. The association with von Hippel-Lindau syndrome was ruled out as magnetic resonance imaging brain, computerised tomography abdomen and 24-hour vanillylmandelic acid were all normal. She was given two doses of intravitreal bevacizumab (IVB) following which the serous detachment resolved (Figure 3c). When the edema recurred again, she was given a dose of 4mg intravitreal triamcinolone acetate (IVTA). The edema resolved and did not recur till the end of three months. She was given the option of PDT, however, she did not consent for the procedure due to the high costs involved.

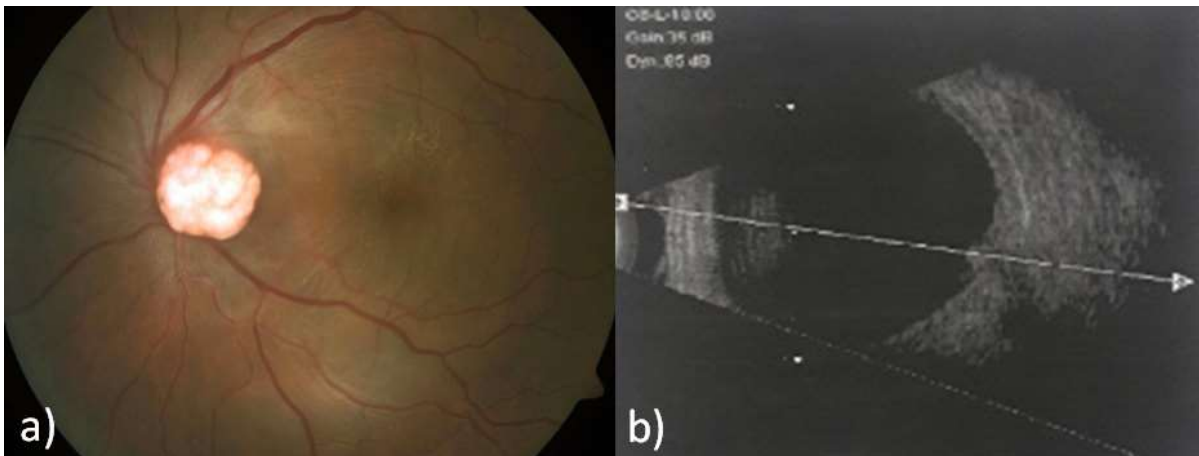


Figure 1: a) Fundus photo showing a well-defined yellowish-red lesion overlying the disc, completely obscuring its view and associated with serous macular detachment, b) B scan showing an acoustical solid mass with smooth anterior border and variable medium reflectivity

Discussion

Papillary RCH doesn't pose a diagnostic challenge. [6] However, due to their anatomical location, they are definitely a therapeutically challenge. Techniques like feeder vessel coagulation and cryotherapy, which are easily applicable to peripheral lesions cannot be considered an option for such lesions. They have a progressive course and with the time, irrespective of growth, the vascular walls become incompetent causing leakage and subsequently macular edema. [1-5]

We presented a sporadic case of solitary endophytic papillary RCH presenting with serous detachment. Although the edema responded initially to IVB, it recurred soon. Intravitreal injections can be used as a temporary remedy for the treatment of macular edema caused by RCH. [5] Definitive and ideal permanent treatment of this condition is still a dilemma. [3,4] Garcia-Arumi *et al* reported that argon laser can effectively treat the lesions. However, it can result in arcuate scotoma. [3] Schmidt-Erfurth *et al* first reported the use of PDT for the

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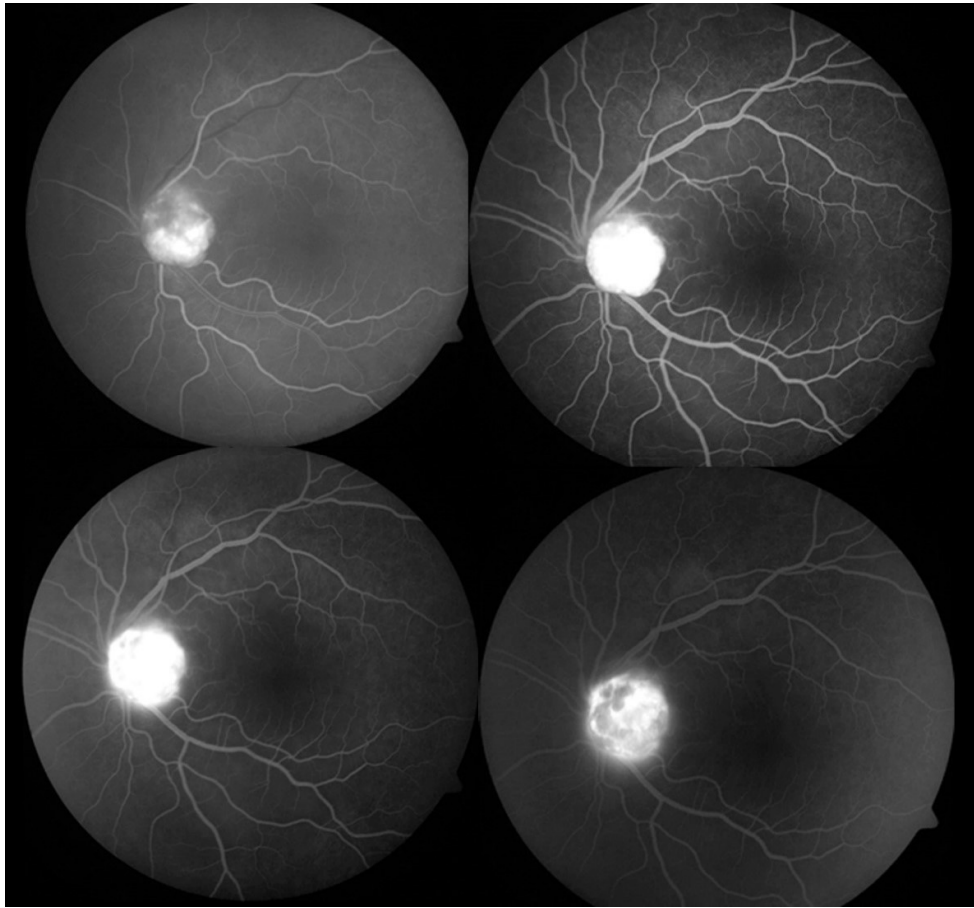


Figure 2: Fundus fluorescein angiography showing early hyperfluorescence, gradually increasing intensity in mid-phase and gradually decreasing intensity in late phase

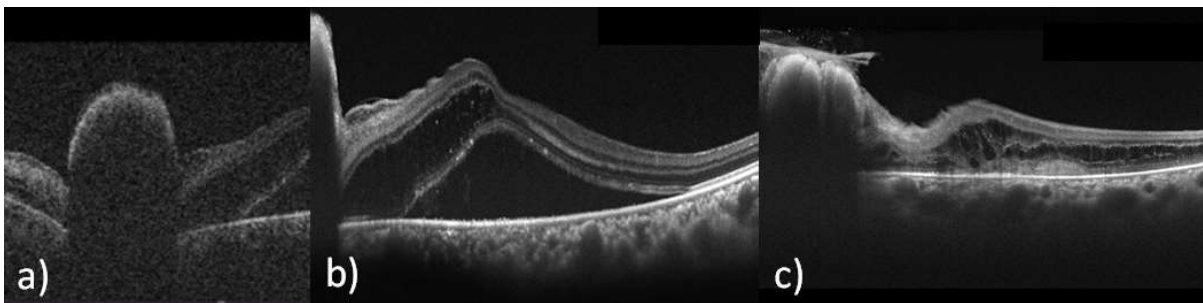


Figure 3: Optical coherence tomography images showing a) hyperreflective mass overlying the disc, b) serous retinal detachment at the macula, and c) resolution of serous detachment after two doses of intravitreal bevacizumab.

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treatment of papillary RCH. They reported that PDT can achieve adequate shrinkage and fibrosis of the lesion. However, it was associated with a high rate of retinal venous occlusion and optic nerve ischemia.⁴ Navea-Tejerina *et al* reported increase in traction over retina that required vitrectomy after the use of PDT for the treatment of papillary RCH.⁷ The successful use of PDT for the treatment of papillary RCH without causing any complications remains limited to mere case reports.⁸

Currently, there is no ideal definitive treatment available for papillary RCH. Larger studies are necessary to establish the treatment for this condition. However, this is a very rare disease and such trials are difficult.

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