Scholars Journal of Medical Case Reports (SJMCR) **ISSN 2347-6559 (Online)**
Abbreviated Key Title: Sch. J. Med. Case Rep. **ISSN 2347-9507 (Print)**

A United of Scholars Academic and Scientific Society, India

Solitary Retinal Capillary Haemangioblastoma: A Case Report

Diymitra K. Ganasan[1*], Mushawiahti Mustapha[2], Aida Zairani Mohd Zahidin[3]
[1]Jabatan Oftalmologi, Tingkat 9, Pusat Perubatan Universiti Kebangsaan Malaysia, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Batu 9 Cheras, Wilayah Persekutuan Kuala Lumpur
[2]Jabatan Oftalmologi, Tingkat 9, Pusat Perubatan Universiti Kebangsaan Malaysia, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Batu 9 Cheras, Wilayah Persekutuan Kuala Lumpur
[3]Jabatan Oftalmologi, Tingkat 9, Pusat Perubatan Universiti Kebangsaan Malaysia, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Batu 9 Cheras, Wilayah Persekutuan Kuala Lumpur

***Corresponding author**
Diymitra K. Ganasan

Article History
Received: 13.04.2018
Accepted: 22.04.2018
Published:30.04.2018

DOI:
10.36347/sjmcr.2018.v06i04.011

Abstract: Retinal capillary hemangioblastomas (RCH) are usually associated with von Hippel-Lindau (VHL) disease although they may present as a solitary tumour without systemic involvement. We present a case report on a healthy young female who is found to have a solitary retinal capillary hemangioblastoma. The presenting ocular complaint was floaters and on examination vision was 6/6. Clinical findings and angiographic studies supported the diagnosis. She was initially observed as her vision was good and there was no macula involvement. Unfortunately she suffered a drop in visual acuity and was then subjected to five intravitreal Ranibizumab injections and cryotherapy. She managed to retain best corrected visual acuity (BCVA) of 6/9 following treatment. This case report is to highlight the unusual occurrence of solitary retinal capillary haemangioblastoma, various possible differential diagnoses to be considered and the treatment options available.
Keywords: retinal capillary haemangioblastoma, von Hippel Lindau.

INTRODUCTION

Retinal capillary haemangioblastoma (RCH) is one of the presenting features of von Hippel Lindau (VHL) disease and may lead to visual impairment if undetected [1]. Approximately 50% of patients found to have solitary RCH are expected to be diagnosed with VHL disease [2].

However the risk of diagnosis reduces with increasing age. We present a case report on a young female with solitary RCH who presents with floaters as the only ocular complaint. The possible differential diagnoses and treatment are outlined as below.

CASE REPORT

A 31 year old lady who was premorbidly well presented to the Eye Casualty clinic with the complaint of left eye floaters. The floaters had been present for 5 years and had not been worsening. There were no flashes of light, blurring of vision, eye pain, eye redness, tearing or discharge. There was also no history of trauma.

On ocular examination, visual acuity (VA) for both eyes were 6/6, N6. There was no relative afferent pupillary defect. Anterior segment examination and intraocular pressures were normal bilaterally. The left eye fundus showed a retinal capillary haemangioblastoma in the superotemporal quadrant supplied by two dilated and tortuous retinal veins. There was no surrounding subretinal fluid seen. At the macula there was an epiretinal membrane and presence of collaterals. Otherwise, there were no retinal exudates or vitreous haemorrhage. Right eye fundus and systemic examination were unremarkable.

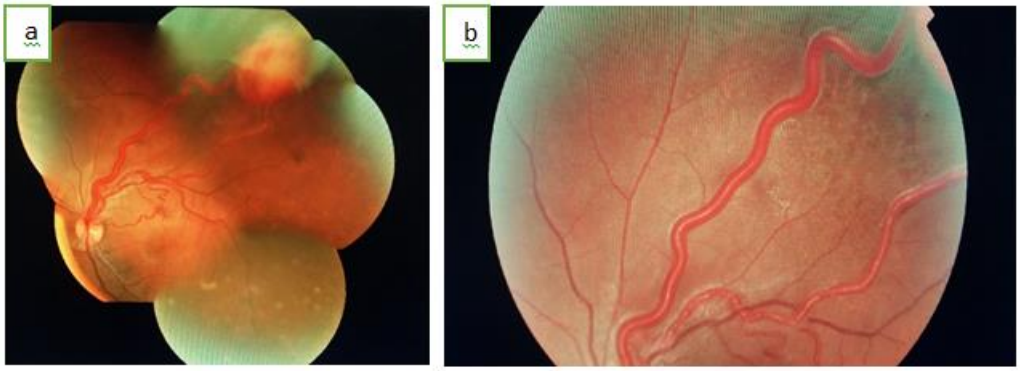

Fig-1 [a,b]: Fundus picture shows retinal capillary haemangioblastoma and feeder vessels

Optical Coherence Tomography (OCT) macula done showed minimal subretinal edema superiorly, not involving the fovea. Fundus fluorescein angiography (FFA) showed leakage and presence of feeder vessels. We scheduled a Magnetic Resonance Imaging (MRI) scan of Brain and Ultrasound Abdomen for her in which results were normal. She was given monthly follow-up during which her vision and condition remained stable.

Five months later during her clinic visit, we noted her best corrected visual acuity (BCVA) dropped to 6/18 over the affected eye. She did not have any new symptoms and clinical examination revealed no increasing macula oedema, exudates or retinal detachment. Serial OCT macula showed stable minimal subretinal fluid not involving fovea. In view of her drop in VA she was treated with a total of five intravitreal Ranibizumab injections. However after the third injection, subretinal fluid level remained the same. She then underwent left eye anterior cryopexy following which repeat FFA showed reduction in leakage. Two weeks later vision improved to 6/12 pinhole 6/9. OCT macula one week post procedure showed further reduction in subretinal fluid.

DISCUSSION

Von Hippel-Lindau (VHL) disease is a familial cancer syndrome characterized by multi systemic tumours and cystic lesions. [1] When our patient presented to the Eye Casualty with a retinal capillary haemangioblastoma (RCH), one of the top differentials were von Hippel Lindau (VHL) disease. This is as RCH is the most common and earliest presentation of von Hippel Lindau disease. [2] The incidence of RCH in VHL disease has been reported to range from 49% to 85%. [3] This patient was screened for VHL disease via Ultrasound Abdomen and Magnetic Resonance Imaging (MRI) brain to look out for intracranial haemangioma, renal cell carcinoma, pheochromocytoma and pancreatic cysts. [3] However this patient was found to only have a solitary RCH and there was no evidence of systemic disease. Few criteria that suggest VHL disease are multiple retinal RCH lesions (2 or more), other manifestations of VHL disease or positive family history [2].

This patient presented not far off from the average age of onset of 25 years [1]. However she presented with a unique complaint of floaters instead of blurring of vision. This is likely due to the peripheral location of the tumour which was not significant enough to cause visual impairment. In certain cases, peripheral tumours may also lead to decrease in vision if there is significant increase in size, increase in vascular permeability (leading to accumulation of subretinal fluid) and presence of hard exudates at macula [4]. The floaters were attributed to the minimal vitreous condensation seen in the fundus.

A few differentials for a RCH would include retinal microaneurysm or Coats disease. In a retinal microaneurysm there would be round or fusiform dilatation of retinal arteriole usually from a branch of the supero-temporal vessels [5]. In the latter, the age of presentation is during childhood and the abnormal vasculature is seen more diffusely [3]. Another differential would be retinal cavernous haemangioma however there would not be prominent feeder vessels as seen in our patient. Meanwhile, vasoproliferative tumours may present in a similar manner but are commonly found at the inferior quadrant and lack feeder vessels [3].

Treatment of retinal capillary haemangioblastoma is varied as it largely depends on its location, size and associated complication [1]. In this patient we initially observed her first as her vision was good and there was no involvement of the fovea. Active surveillance is recommended for those with peripheral tumours < 500um in size with no exudation [1]. After her vision reduced to 6/18 unaided, we administered intravitreal Ranibizumab injections in an effort to try and reduce the size of the haemangioblastoma. Intravitreal anti-vascular endothelial growth factors (VEGF) are thought to be useful in treating RCH due to poor regulation of VEGF in the disease [7]. In a study done by Wong et al, intravitreal Ranibizumab injections done every four weeks delivered minimal beneficial effects [6]. However it was admitted that future studies were needed to determine the definitive role of anti-VEGF in treatment of RCH [6].

After five injections of Ranibizumab, our patient was treated with cryotherapy. Based on a study done by Atik *et al*. cryotherapy is thought to result in reduction of subretinal fluid [1]. Indications for cryotherapy included large peripheral tumours more than 3mm in size with subretinal fluid as seen in our patient [1,3]. Other options of treatment are laser photocoagulation which is usually reserved for small RCH located in the posterior pole in eyes with clear media [3].

CONCLUSION

In conclusion, solitary RCH are uncommon findings in ophthalmology and due treatment is indicated when necessary to preserve visual outcome.

REFERENCES

1. Atik SŞ, Solmaz AE, Öztaş Z, Eğrilmez ED, Uğurlu Ş, Atik T, Afrashi F. Von Hippel-Lindau disease: The importance of retinal hemangioblastomas in diagnosis. Turkish journal of ophthalmology. 2017 Jun;47(3):180.
2. Singh A, Ahmad N, Shields C, Shields J. Solitary retinal capillary hemangioma: Lack of genetic evidence for von Hippel-Lindau disease. Ophthalmic Genetics. 2002;23(1):21-27.
3. Von Hippel-Lindau Disease. Available from http://retinatoday.com/2010/06/von-hippel-lindau-disease
4. Chittiboina P, Lonser R. Von Hippel–Lindau disease. Neurocutaneous Syndromes. 2015;:139-156.
5. Retinal Capillary Hemangioblastoma and von Hippel-Lindau Disease. Available from http://eyewiki.aao.org/Retinal_Capillary_Hemangioblastoma_and_von_Hippel-Lindau_Disease
6. Wong WT, Liang KJ, Hammel K, Coleman HR, Chew EY. Intravitreal ranibizumab therapy for retinal capillary hemangioblastoma related to von Hippel-Lindau disease. Ophthalmology. 2008 Nov 1;115(11):1957-64.
7. Pinarci E, Karacal H, Demirel B. Intravitreal Bevacizumab Followed By Laser Photocoagulation for Retinal Capillary. Retinal Cases & Brief Reports. 2012;6(1):76-79.