

Case Report

Vitreous hemorrhage as a clue to late presenting Pulmonary Arterio Venous Malformation



Brijesh Takkar^{*}; Archita Singh; Prateek Kakkar; Shorya Azad; Rajvardhan Azad

Abstract

Retinal vasculature is frequently altered in systemic vascular disorders such as Diabetes and Hypertension. We present a rare case in which vitreous hemorrhage was the presenting sign of an underlying congenital cyanotic heart disease.

Keywords: Vitreous hemorrhage, Cyanotic heart disease, Conjunctival telangiectasia

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Introduction

Retinal vasculature changes are well documented in systemic diseases such as Diabetes and Hypertension. Although proliferative retinopathy has been described in cases of Congenital Cyanotic Heart Disease (CCHD),¹ we discuss a unique case in which Vitreous Hemorrhage (VH) led to diagnosis of Pulmonary Arterio Venous Malformation (PAVM).

Case report

A 21-year-old male presented to the outpatient department with loss of vision in left eye (LE) which was sudden in onset. He denied any history of trauma, recurrent bleeds or other systemic disease. He was evaluated at a local hospital when the eye symptoms first started 2 months back and was diagnosed as having Eales' Retinal Vasculitis with VH. He was treated with retinal laser twice without any clinical or photographic documentation of vasculitis related changes in either eye or on systemic examination. Family history was not significant. Ocular Examination revealed best corrected visual acuity (BCVA) of 6/6 and 1/60 in right eye (RE) and left

eye (LE) respectively. While IOP, ocular adnexa and anterior Segment of both the eyes appeared to be normal, LE fundus examination revealed fresh VH. Minimal tortuosity of peripheral retinal vessels was detected in RE. To our surprise, general physical examination revealed central cyanosis and clubbing which on leading questions were found to have been present since early childhood. At this stage ocular examination was repeated and a very small patch of Bulbar Conjunctival Telangiectasia (BCT) was detected near the superior limbus of RE, previously hidden by the upper eye lid (Fig. 1). The patient was unaware of these findings of BCT. Fluorescein angiography revealed no leaks/hyperfluorescence in RE. Retina was found to be attached on sonography of LE. Hence a presumptive diagnosis of Sickle Cell Retinopathy was made and further investigations ordered.

Blood investigations revealed hemoglobin to be 18.6 gm %, a high RBC count of 7.46 million/cubic-mm and raised packed cell volume (60.8%). Total leukocyte count, peripheral blood smear, hemoglobin chromatography (for sickle cell retinopathy), bleeding time and clotting time were within normal limits. Chest X-ray was ordered which revealed cardiomegaly. Patient was then immediately referred to a

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Dr. Rajendra Prasad Centre for Ophthalmic Sciences, AIIMS, New Delhi 110029, India

^{*} Corresponding author at: Vitreo-Retina Services, Dr. R P Centre, AIIMS, New Delhi, India. Fax: 01126593101. e-mail addresses: britak.aiims@gmail.com (B. Takkar), architasingh88@gmail.com (A. Singh), prateek.kakkar.10@facebook.com (P. Kakkar), shoryaazad@hotmail.com (S. Azad), rajvardhanazad@hotmail.com (R. Azad).



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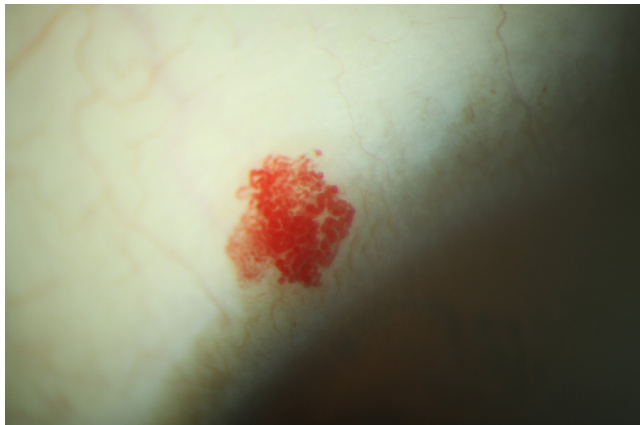


Figure 1. Clinical photograph demonstrating Bulbar Conjunctival Telangiectasia at the limbus.

cardiologist where echocardiography revealed moderate left ventricular dysfunction (ejection fraction of 30–35%) with Right Pulmonary Artery to Left Atrium Fistula. Computerised tomography angiogram confirmed the fistula with aneurismal left atrium and right lower lobe pulmonary venous varix. A fistula ligation procedure was done for the same.

The patient was reevaluated by us after systemic optimization and amelioration of cyanosis. While the BCT persisted, peripheral retinal vessel tortuosity had decrease in RE and the LE VH appeared to be old and laser marks were visible. There was no evidence of any recurrence or fresh VH. The patient was taken up for LE 25 G Pars Plana Vitrectomy during which partial separation of the posterior hyaloid was noted along with 2 peripheral fibrotic neovascular fronds which were diathermised and trimmed. No peripheral vascular tortuosity, vascular sclerosis or any other sign of retinal vascular occlusion/vasculitis was seen. One month later patient had a UCVA of 6/9 and IOP of 12 mmHg in LE along with attached retina, no epiretinal membranes while a trimmed fibrous frond was seen at the optic disc (Fig. 2).



Figure 2. Postoperative fundus photograph. Laser marks are visible below inferior arcade along with a trimmed fibrous frond at the optic disc.

RE BCT still persisted. After careful literature search the patient was labelled as a suspect case of Hereditary Hemorrhagic Telangiectasia (HHT)² and advised routine follow-up and cardiac care. Ocular and cardiac condition were stable at 3 months of follow-up.

Discussion

This case highlights several important issues. VH can have variable causes and although Eales' disease is very rampant in the Indian subcontinent,³ it should only be a diagnosis of exclusion especially in the absence of retinal inflammation. Young patients may ignore mild symptoms and therefore tailored workup should be ordered based on findings.

To diagnose HHT at least 3 out of 4 (recurrent epistaxis, family history, visceral AVMs and mucocutaneous telangiectasias) criteria must be met, hence our case could only be labelled as a suspect. Neovascular fronds have been rarely described in CHD/HHT^{1,4} children with a proven diagnosis of heart disease. In a very old report, vitrectomy has been described for haemorrhages in known cases of HHT.⁵ To the best of our knowledge VH has not been described at presentation or as the presenting cause of PAVM or in HHT. In fact chorioretinal atrophy is believed to be a common posterior segment manifestation of HHT⁶ with retinal telangiectasia only in around 1% of cases⁷.

Proliferative retinopathy can occur in CHD due to chronic lack of oxygen in the arterial blood leading to hypoxia.^{1,4} Another possible cause for proliferation could be hyperviscosity syndrome due to polycythemia as evidenced by tortuous retinal vessels in RE. The absence of such vessels was noted during surgery in LE but, as seen in RE, it may have happened due to reversal of hypoxia after cardiac surgery. Vessels in HHT are known to be fragile with poor contractility,⁸ hence prone to rupture and bleeding. In the absence of sclerosed vessels, inflammation and hypertension, old vascular occlusions are very unlikely although arterial occlusion has been described in HHT.

BCT, often misdiagnosed as episcleritis, is known to be present in ophthalmic conditions such as ocular rosacea and post radiation therapy as well as a large number of systemic conditions including HHT, Bloom Syndrome, Louis Barre syndrome, Essential Telangiectasia, Alport Syndrome and Sickle Cell retinopathy. BCT (Fig. 1) of our patient remained unchanged in size during his treatment unlike the tortuous peripheral retinal vessels. Hence BCT was independent of the PAVM favouring the diagnoses as HHT suspect rather than Idiopathic PAVM.

VH can therefore be the presentation of a serious underlying cardiac disorder and such cases should undergo careful evaluation. The presence of BCT mandates high suspicion and careful workup. Visual prognosis in such cases of CHD appears to be favorable in the absence of complications.

Conflict of interest

The authors declared that there is no conflict of interest.

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