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Acute Vision Loss in a Patient with Chronic Esophageal Stenosis

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Abstract

Purpose: To describe a unique cause of Valsalva Retinopathy (VR) with an alternative surgical approach to chronic non-clearing pre-foveal hemorrhage.

Method: Case presentation.

Results: A 45-year-old African American female presented with acute vision loss from 20/20 to count fingers (CF) in her right eye. Ophthalmoscopy and ocular coherence tomography (OCT) showed old yellow-red pre-retinal opacity, in the sub-hyaloid and sub-internal limiting membrane (ILM) planes, obscuring the fovea. After 1 month of conservative management with no improvement, Pars Plana Vitrectomy (PPV) with posterior hyaloid membrane removal without ILM peeling was performed with the patient's best-corrected visual acuity (BCVA) dramatically improved to 20/25.

Discussion: We raise the question regarding the role of ILM peeling in treating premacular hemorrhage. Even without ILM peeling, our patient's hemorrhage resolved after the procedure. This suggests that PPV combined with posterior hyaloid removal is a safer and effective alternative to surgical treatment in patients with certain clinical conditions. In addition, we provide clear evidence to support the location of the hemorrhage in VR as both sub-hyaloid and sub-ILM.

Keywords: Choking, ILM peeling, Premacular hemorrhage, Valsalva retinopathy

Abbreviations: VR: Valsalva Retinopathy; CF: Count Fingers; OCT: Ocular Coherence Tomography; ILM: Internal Limiting Membrane; PPV: Pars Plana Vitrectomy; BCVA: Best-Corrected Visual Acuity; OD: Oculus Dextrus; DD: Disc Diameter; PVD: Posterior Vitreous Detachment; SCR: Sickle Cell Retinopathy; Nd: YAG - Neodymium-doped Yttrium Aluminum Garnet

Introduction

The Valsalva maneuver is a common action performed during activities such as coughing, sneezing, vomiting, straining, and heavy lifting. It is characterized by exhalation against a closed glottis resulting in increased intrathoracic and intraabdominal pressures, which raise pressures in the valveless venous system. Valsalva Retinopathy (VR) is a complication of increased intraocular venous pressure resulting in subsequent rupture of superficial retinal capillaries with pre-retinal, retinal or vitreous hemorrhages, the location of which has classically been debated [1].

We present a patient who developed acute painless vision loss due to retinal hemorrhage in association with a severe acute choking episode secondary to chronic esophageal stenosis.

Case Presentation

A 45-year-old African American female presented with acute loss of vision in her right eye 6 weeks prior to presentation. She stated that she went to bed with normal
vision and woke up the next morning with vision loss and an accompanying red tint of vision oculus dextrus (OD). Her past medical history included anemia with an unsubstantiated diagnosis of either beta-thalassemia or sickle cell, as well as esophageal stenosis. She denied any pain, recent trauma, heavy lifting, or coughing prior to losing her vision. Her previous BCVA was 20/20 but at her initial examination, she was Counting Fingers (CF) at 5ft, right eye and 20/20, left eye. The intraocular pressure was 12 mmHg, right eye, and 14 mmHg, left eye. The anterior segment was normal.

Ophthalmoscopy showed a well-circumscribed 1 disc diameter (DD) old yellow pre-retinal hemorrhage obscuring the fovea OD (Figure 1).

Initial optical coherence tomography (OCT) demonstrated similar findings of a fovea obscured by likely old sub-hyaloid and sub-internal limiting membrane (ILM) hemorrhage (Figure 2).

**Figure 1:** Initial color fundus photograph demonstrating old yellow sub-hyaloid hemorrhage OD.

**Figure 2:** Initial OCT image of our patient demonstrating hemorrhage OD.

History and clinical examination at this time supported a differential of Hemorrhagic Posterior Vitreous Detachment (PVD), Sickle Cell retinopathy (SCR), Beta-Thalassemia retinopathy, Trauma, or VR. However, the patient’s denial of trauma/straining and lack of PVD on exam made SCR or Beta-Thalassemia more likely diagnoses. The patient was observed for 1 month without improvement in fundus examination or visual acuity. In addition, Fluorescein Angiography did not demonstrate neovascularization. The patient was scheduled for a Pars Plana Vitrectomy (PPV) due to non-clearing central hemorrhage and continued poor vision. As part of pre-op clearance for anemia, she established care with Hematology and was found to be severely iron deficient. The type of her hemoglobinopathy remained unclear due to her severe iron deficiency, but a sickle component to her anemia was ruled out. We believe that the majority of the hemorrhage was trapped in sub-hyaloid space. As a result, the surgical plan was to induce PVD with Kenalog staining and remove the posterior hyaloid membrane and hemorrhage. Intraoperatively, there was a small piece of hyaloid membrane above the fovea that was difficult and too dangerous to remove, possibly due to the adhesion effect from the sub-hyaloid hemorrhage. As part of the
plan, the ILM was not peeled. Scleral depression did not demonstrate evidence of peripheral neovascularization or vasculopathy. After vitrectomy, the size and color of the hemorrhage reduced remarkably (Figure 3).

1-week postoperatively, the patient's BCVA was dramatically improved to 20/25 and serial fundus examination/OCT demonstrated a small amount of residual Sub-ILM blood decreasing over time (Figure 4).

The etiology of this patient's retinal hemorrhage was still uncertain; however, after further review of her chart, it was discovered that the patient's esophageal stenosis was severe enough to warrant monthly balloon dilatations due to symptomatic choking. Upon further inquiry, the patient recalled a significant choking episode the day preceding her vision loss, so severe that she considered calling an ambulance.

Hence, we report the first case of VR caused by severe choking secondary to esophageal stenosis.

**Discussion**

VR is characterized by rupture of superficial retinal capillaries. This can lead to extravasation of blood below the ILM and/or sub-hyaloid space. It often occurs in healthy young adults and has been associated with a large variety of activities from aerobic exercises, push-ups, constipation, pregnancy, labor, playing a wind instrument, sexual activity and choking [1-3].

VR typically has a good prognosis with complete recovery back to baseline vision within weeks to months after onset, as a large proportion of hemorrhages resolve on their own especially when less than 1 DD [4]. Surgical interventions may be indicated with non-clearing vitreous and retinal hemorrhages as permanent visual impairment may result from toxic damage to the retina caused by extended exposure to persistent hemoglobin and iron. Techniques to treat premacular hemorrhage include using a neodymium-doped yttrium aluminum garnet (Nd-YAG) to puncture the posterior hyaloid face, intravitreal tissue plasminogen activator with pneumopexy, and PPV [4,5].

Nd-YAG involves the opening of the vitreoretinal interface and releasing the entrapped blood into the vitreous cavity. This technique is most useful in the treatment of non-dense premacular hemorrhages in the sub-hyaloid, less than 3 weeks old, and larger hemorrhages, typically > 3 DD. However, when premacular blood is coagulated, drainage with YAG laser
is hindered. In addition, possible severe complications of Nd: YAG laser include retinal detachment, epiretinal membrane formation, and the creation of a macular hole or persistent premacular cavity [4,6,7].

ILM peeling refers to the surgical technique for the removal of preretinal tissue and ILM in the macula and is always performed in conjunction with PPV. While the main indications for PPV are epiretinal membranes and macular holes, it is the most common surgical option for non-clearing vision-threatening hemorrhage in VR. For the past 20 years, there has been controversy regarding the role of ILM peeling in vitreoretinal surgery, altering the structure of the retina is not without risk. Complications include decreased retinal sensitivity, microscotomas, iatrogenic macular holes, and focal retinal hemorrhages and edema [8]. The exact plane of retinal hemorrhage in VR may be difficult to determine, especially in the absence of PVD [6]. The location of our patient’s hemorrhage was thought to be both sub-hyaloid and sub-ILM based on the initial OCT (Figure 2). In our case, patient’s hemorrhage resolved with PPV and posterior hyaloid membrane removal without ILM peeling (Figures 3-5). There was a small amount of sub-ILM hemorrhage still present post-operatively that resolved over time (Figures 3-5).

Figure 5: Color Fundus photographs (A) and OCT (B) Post-Op month 3.

This suggests that in select patients, withholding ILM peeling or PPV alone to remove vitreous will enhance the resolution of hemorrhage remarkably and may be a safer and equally effective approach to the ILM peeling surgical treatment. Further research is needed to establish a clear guideline on when PPV with ILM peel is performed and when ILM peel can be withheld.

Lastly, we believe our case provides clear pre-operative and post-operative evidence that the hemorrhage in VR is both sub-hyaloid and sub-ILM.

Declarations

The authors are responsible for the writing of the article. Each author contributed significantly. The authors have no conflicts of interest. Funding for this work provided by the Temple University Hospital, Department of Ophthalmology. Consent was not required.

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