Valsalva retinopathy in the third trimester of pregnancy

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Case Details
MD, a 24-year old female of African descent, presented upon urgent referral from her family physician with an 18-hour history of a single central floater in her left eye not accompanied by pain or photopsia. A vague occipital headache was reported. There was no history of trauma. Her general health was good, and no systemic medications were being taken. However, MD was in her third trimester of pregnancy, and reported an episode of violent vomiting immediately preceding the onset of symptoms.

Entering unaided visual acuities were 6/7.5+ in the right, 6/7.5- in the left. Ocular motility was full.

Confrontation visual field assessment was full to finger counting in both right and left. Pupil reactions were normal with no afferent defect. Anterior segment evaluation was unremarkable; there was no neovascularization. Intraocular pressures were 12 and 13, right and left respectively, at 10:00 a.m.

Dilated retinal examination of the pole and periphery (using my standard protocol of 0.5% tropicamide followed by 60 seconds of gentle eyelid closure) was unremarkable in the right eye (Figure 1). In the left, a predominantly inferior-central vitreous hemorrhage (VH) with several superior ‘streamers’ was noted (Figure 2). There was no obvious neovascular process, or macular or peripheral retinal anomaly.

Keywords: Valsalva maneuver, Valsalva retinopathy, hyaloidotomy, Terson’s syndrome, Purtscher retinopathy

Figure 1 – healthy posterior pole, right eye

Figure 2 – Valsalva retinopathy, left eye

ABSTRACT | RÉSUMÉ

MD, a 24-year old female of African descent in her third trimester of pregnancy, presented upon referral from her family physician with an 18-hour history of dark floating spot in her left eye. Her history was significant for violent vomiting immediately preceding the onset of visual symptoms. General and ocular health history was otherwise unremarkable. Clinical examination showed central vitreous hemorrhaging in the absence of any other ophthalmic anomaly, consistent with a diagnosis of Valsalva retinopathy. Over the subsequent several months, which included the uncomplicated birth of her healthy child, the hemorrhaging resolved without sequelae.

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MD, femme de 24 ans de descendance africaine qui en est au troisième trimestre de sa grossesse, se présente recommandée par son médecin de famille en se plaignant d’avoir un point flottant noir depuis 18 heures dans l’œil gauche. Comme antécédents importants, elle a vomi violemment juste avant l’apparition des symptômes visuels. Ses antécédents relatifs à la santé générale et oculovisuelle ne présentaient autrement rien de remarquable. L’examen clinique révèle une hémorragie au centre du vitré et aucune autre anomalie ophthalmique. Ce qui concorde avec un diagnostic de rétinopathie de Valsalva. Au cours des mois qui suivent, qui ont inclus la naissance sans complication de son enfant en bonne santé, l’hémorragie s’est résorbée sans laisser de séquelles.

Mots clés: Manœuvre de Valsalva, rétinopathie de Valsalva, hyaloidotomy, syndrome de Terson, rétinopathie de Purtscher
Given that the patient had been examined by her family physician earlier the same day, systemic blood pressure was not reassessed. In the absence of posterior vitreous detachment (PVD) and contributing systemic and/or ophthalmic disease a working diagnosis of Valsalva retinopathy was established. A report was forwarded to the family physician suggesting further investigation for systemic vasculopathies including hypertension and blood dyscrasias. No reply was received.

At follow-up in two weeks MD noted a smaller more peripheral spot in the left eye not accompanied by pain or photopsia. Unaided acuities were 6/6 in the right, 6/6+ in the left. Confrontation visual field assessment remained full; pupil reflexes remained normal with no afferent defect.

Dilated retinal examination revealed significant resolution of the VH, with some ‘greying’ of the remaining blood (Figure 3). The macula and peripheral retina remained unremarkable.

As the patient was approaching her due date, further follow-up was scheduled for three months, and an update was forwarded to the family physician.

Four months following the onset of her symptoms, MD reported a small persistent floater in the left eye. No exacerbation of her symptoms had occurred during the uncomplicated vaginal birth of her healthy child 9 weeks earlier. Unaided acuities remained 6/6 in the right, 6/6- in the left. Confrontation visual field assessment remained full; pupil reflexes remained normal with no afferent defect.

Dilated retinal examination revealed essentially complete resolution of the VH in the left eye (Figure 4). The macula and peripheral retina remained unremarkable. An update was forwarded to the family physician.

Thirteen months following onset of her symptoms, MD noted no floater, eye pain, or photopsia in either eye, but slight near strain and occasional headache. Best-corrected acuities were 6/6 in both right and left (O.D. +1.25-0.25×180; O.S. +1.00-0.25×180). Binocular vision assessment indicated a 4Δ near exophoria. Motility was full. Confrontation visual field assessment remained full; pupil reflexes remained normal with no afferent defect. Anterior segment examination was unremarkable. Intraocular pressures were 11 and 13, right and left respectively, at 4:40 p.m.

Dilated retinal examination revealed no anomalies of the pole or periphery of the right eye (Figure 5). The left pole and peripheral retina were unremarkable, while the inferior vitreous showed trace hazing consistent with resorbed VH (Figure 6).

Annual comprehensive examinations were recommended.
Discussion

The Valsalva maneuver, named for the Italian anatomist who first investigated it in the 17th century, is defined as forcible exhalation effort against a closed glottis causing a sudden and significant increase in intrathoracic pressure. The resulting compression of the thoracic vena cava causes an increase in intravenous pressure that interferes with venous return to the heart.

Given that veins above (rostral to) the heart have no valves, relying primarily upon gravity for venous return, this elevated pressure is readily transferred to the ophthalmic venous system. Intravenous pressure, if elevated to a level of 100 mmHg or more, can damage ophthalmic capillaries, causing hemorrhaging. A history of recent Valsalva maneuver is helpful in establishing a diagnosis: vigorous sexual activity, heavy lifting, constipation, roller-coaster riding, childbirth, sneezing, exercise, dancing, gastroenteroscopy, pneumoencephalography, trauma, cardiopulmonary resuscitation, orotactical contests, riding a motorcycle, bungee jumping, and inflating balloons are among a host of interesting and diverse causes.

Hemorrhages can be unilateral or bilateral, single or multiple, of varying shape, and in a number of ocular structures, including the periocular skin, subconjunctival and suprachoroidal spaces, and perifoveal retina. The precise location of a retinal hemorrhage (sub-internal limiting membrane (ILM) versus subhyaloid) can be difficult to determine through ophthalmoscopy, although sub-ILM hemorrhages tend to be more immobile. In the presence of a posterior vitreous detachment (PVD), the initial hemorrhage typically occupies the sub-ILM space, given that there is no attached hyaloid membrane to confine the blood. Hemorrhaging initially occupying the sub-ILM space may pass through ILM breaches into the subhyaloid space, and subsequently through the posterior hyaloid face to involve the vitreous itself.

Intravenous fluorescein angiography (IVFA) will show blocked fluorescence in the area of the hemorrhage. Optical coherence tomography (OCT) allows more precise localization of intraocular hemorrhaging, and has revealed the potential for preretinal membrane (PRM) formation. PRM are histologically similar to the glial tissue of epiretinal membranes (ERM), but contain blood degradation products and are not adherent to the underlying retinal surface.

Symptoms depend upon both the severity and location of the hemorrhage. When the visual axis is involved, vision can be significantly impaired. Given that small extrafoveal hemorrhages may be minimally symptomatic, Valsalva retinopathy is likely under-reported. Fortunately, most cases resolve spontaneously over several weeks to months of observation, with vision often improving to normal or near normal, as was the case with patient MD.

Permanent visual impairment may result from macular pigmentary alterations, fibrotic ERM/PRM formation, or toxic retinal damage secondary to prolonged exposure to blood, hemoglobin, and/or iron, in increasing order of toxicity. Large sub-ILM or subhyaloid hemorrhages not resolving spontaneously may be evacuated into the vitreous space by performing a posterior vitreous hyaloidotomy using the Nd:YAG laser. This treatment allows clearing of persistent hemorrhage and commensurate improvement in acuity within 14 to 28 days in up to 90% of cases, versus a mean of nearly 50 days for patients simply observed. Given that patients with Valsalva retinopathy are often healthy young adults who respond particularly well to this treatment providing that the blood has not started to clot, a speedy visual recovery is certainly desirable.

That being said, hyaloidotomy is not without potential iatrogenic complications including peripheral retinal break, macular hole, ILM wrinkling, and ERM formation. It is, however, less invasive than vitrectomy, and greatly reduces the risk of potentially devastating infectious complications. Subsequently or alternatively, vitrectomy may still be necessary in cases of severe or slowly resolving vitreous hemorrhage (VH). Following vitrectomy, residual sub-ILM hemorrhage may be released through ILM peel, or laser membranotomy. Subsequent histological evaluation has shown excised ILM to be convoluted and anomalous, containing atypical cellular and blood degradation components. An element of inflammatory ‘proliferative vitreoretinopathy-like’
damage may be responsible for some cases of poor post-event vision, supporting earlier and more aggressive intervention.

Pregnancy has been identified as a predisposing factor in the development of Valsalva retinopathy. Pregnancy-induced hormonal, immunologic, metabolic, and hematologic changes including thrombocytopenia can increase the likelihood of both Valsalva maneuver and retinopathy. Constipation and nausea, known triggers of Valsalva maneuver, frequently accompany pregnancy. Elevation of intra-abdominal pressure secondary to uterine expansion can significantly increase ‘baseline’ intravenous pressure, exacerbating any further increase occurring during labor and delivery. There are, however, no reports detailing recurrent Valsalva retinopathy following vaginal delivery, making additional accommodation unnecessary. Retinal hemorrhaging has been reported in association with both epidural anesthesia, typically required for elective Caesarean section, and general anesthesia. Both trigger an increase in venous pressure, and the latter may induce Valsalva maneuver during extubation.

Intraocular hemorrhaging in newborns was first described in 1861, only eleven years after the introduction of the ophthalmoscope, and has subsequently been reported in up to 40% of vaginal, but only a small fraction of Caesarean deliveries. Trauma during and following delivery is an important consideration: it has been argued that infants are unable to self-generate the increase in intra-thoracic pressure required to trigger Valsalva retinopathy. Consequently and unfortunately, abusive head trauma (AHT, ‘shaken baby syndrome’) must be included in the differential diagnosis for infants and young children presenting with intraocular hemorrhaging, particularly when hemorrhaging is bilateral and involves multiple retinal layers through the entirety of the fundus, suggesting repetitive acceleration/deceleration injury.

While Duane originally postulated that faulty retinal capillaries were responsible for Valsalva retinopathy, the prevalence of this condition in healthy young adults would suggest otherwise. In fact, the debate as to whether ruptured capillaries are abnormal or healthy has largely been resolved in favor of the latter, with most arguing that, by definition, Valsalva retinopathy occurs in the absence of concurrent ocular or systemic disease. However, it can be exacerbated by retinal vascular anomalies, systemic medications, hemorrhagic fevers, and blood dyscrasias. Coagulability is a particularly important consideration: pertinent to this case, some maintain that any patient of African descent who presents with intraocular bleeding has sickle-cell disease until proven otherwise. Systemic hypertension, retinal vein occlusion, and diabetes, particularly proliferative disease, are important considerations in the differential diagnosis. Severe anemia, leukemia, and chronic alcoholism have also been implicated as risk factors for intraocular hemorrhage.

Particular care must be taken to consider potentially life-threatening diagnoses, including Terson’s syndrome and Purtsher retinopathy.

First reported in 1900, Terson’s syndrome is characterized by the presence of subhyaloid and/or vitreous hemorrhage secondary to intracranial subarachnoid hemorrhage (SAH). This condition is found more frequently in women, consistent with the gender difference in prevalence of aneurysmal SAH. Intraocular hemorrhaging is bilateral in approximately 50% of cases, but often asymmetric. While intracranial hemorrhage itself can be life threatening, the presence of concurrent VH, found in up to 40% of cases but likely underdiagnosed, is associated with a 4.8× increased mortality risk and more guarded outcome should the patient survive. It was originally hypothesized that the intraocular blood was subarachnoid blood transmitted through the optic nerve sheath. However, it is now believed that VH results from venous stasis and subsequent intraocular vessel rupture caused by acutely elevated intracranial pressure (ICP). Ocular complications may include macular hole and retinal detachment, although the prognosis for visual recovery in surviving patients is surprisingly good. Patients with Terson’s syndrome frequently complain of ‘thunderclap headache’ and subsequently present to the emergency room comatose secondary to dramatically elevated ICP.

Purtsher retinopathy, initially described in 1910, is an occlusive microvasculopathy (traumatic retinal angiopathy) characterized by cotton wool spots, multiple retinal hemor-
rhages, and Purtscher flecken.66 The latter, polygonal areas of inner-layer retinal whitening thought to be capillary bed infarcts between arterioles and venules surrounded by an adjacent clear zone, are pathognomonic but not universal.57 Retinal lesions are localized to the posterior pole following compressive injuries to the trunk and/or long bone fracture or crush, and are bilateral in up to 60% of cases.58 “Purtscher-like” (non-traumatic) retinopathy has been reported secondary to acute pancreatitis, orthopedic surgery, renal failure, and childbirth.59,60 In all etiologies, the pathogenesis is felt to be microembolism of fat, air, fibrin clots, and/or amniotic materials, or complement-mediated leukocyte aggregation. The peripapillary capillary network has relatively few arteriolar feeds, making it uniquely susceptible to embolic obstruction. Presenting visual acuity may be 6/60 or less with central scotoma; visual symptoms are delayed up to 48 hours post-illness or -trauma.61 Spontaneous visual recovery to the level of 6/12 or better occurs within weeks to several months in the majority of cases, although dramatically reduced presenting acuity due to involvement of the macular arteriolar network is a poor prognostic indicator. There is no proven treatment: the use of systemic steroid and non-steroidal anti-inflammatory agents is variably effective in improving ultimate visual recovery.62

From a biomechanical perspective, the causative force in Purtscher retinopathy is centrifugal (anterior/posterior), whereas that in Valsalva retinopathy is centripetal (superior/inferior).63 As such, concurrent Purtscher and Valsalva retinopathy is rare, although possible in instances of severe chest trauma. In neither condition is direct ocular trauma an etiologic factor.

**Conclusion**

Valsalva retinopathy may be precipitated by a number of very diverse causes, including pregnancy. Fortunately, spontaneous resolution with recovery of normal or near normal vision is the rule in the majority of cases. Unequivocal evidence of antecedent Valsalva maneuver is helpful in establishing the diagnosis. In its absence, differential diagnosis is critical, particularly in light of the potentially devastating consequences of other etiologies of intraocular hemorrhage. Ophthalmic considerations include but are not limited to Terson’s syndrome, Purtscher retinopathy, and proliferative diabetic retinopathy; contribut ing systemic vasculopathies include but are not limited to anemia and leukemia. If concurrent retinal pathology is suspected, or more rapid clearing of the visual axis is necessary, laser hyalidotomy and/or vitrectomy may be considered. Once Valsalva retinopathy has resolved, given its unique etiology and self-limiting course, follow-up is typically through regular comprehensive eye examinations.

**References**


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