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Case Report

Traumatic Hyphema in a Child with Von Willebrand Disease Type 3: A Challenging Case Report

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Abstract

Von Willebrand disease (VWD) is the most common inherited bleeding disorder, affecting almost 1% of the general population. Type 3 VWD is the most severe form, with severe bleeding phenotype that usually presents with mucocutaneous and other serious bleeding. Ocular bleeding, however, is rare in VWD, and hereby we report a three-year-old girl with type 3 VWD who developed traumatic hyphema; an extremely rare presentation. The child presented 2 days after blunt trauma to her left eye with periorbital ecchymosis, subconjunctival hemorrhage, total hyphema, loss of vision together with high intraocular pressure. She was managed conservatively with bed rest, eye shielding, VWF/FVIII concentrate infusion, tranexamic acid and topical treatment for high IOP. Initial improvement was followed by rebleeding with worsening hyphema in addition to new intravitreal hemorrhage. Her management was challenging in view of lack of cooperation, poor compliance with topical treatment, and the need to optimize blood tests for factor VIII and VWF recovery and trough levels. Her serious eye bleeding necessitated long term prophylaxis with VWF/FVIII concentrate. This case highlights the importance of early recognition, timely hemostatic intervention, and coordinated multidisciplinary management to prevent vision-threatening complications.

Keywords: *Von Willebrand Disease; Traumatic Hyphema; Case Report*

Abbreviations

VWD: Von Willebrand Disease; VWF: Von Willebrand Factor; IOP: Intra Ocular Pressure

Introduction

Von Willebrand disease (VWD) is the most common inherited bleeding disorder, affecting approximately 1% of the general population [1]. It results from quantitative or qualitative defects of von Willebrand factor (VWF), a multimeric glycoprotein essential for platelet adhesion and stabilization of factor VIII [2]. VWD is classified into type 1 and 3, which involve quantitative deficiencies of VWF, as well as types 2A, 2B, 2M, and 2N, which are variants of qualitative defects [3,4].

Among these, type 3 VWD represents the rarest and most severe form, inherited in an autosomal-recessive pattern, characterized by almost complete absence of VWF with markedly reduced factor VIII activity. This profound haemostatic defect disrupts both primary

and secondary hemostasis leading to a marked bleeding tendency even after minor trauma. It typically presents early in life with severe mucocutaneous and musculoskeletal bleeding and a markedly increased risk of life-threatening events, including intracranial and gastrointestinal hemorrhage [5-7].

Ocular hemorrhage is an uncommon manifestation of VWD [8]. Hyphema, defined as the accumulation of blood within the anterior chamber of the eye, is most often trauma-related but may rarely occur spontaneously in severe coagulopathies [9]. While generally self-limited in healthy individuals, hyphema in patients with bleeding disorders can be sight-threatening. Potential complications include rebleeding, raised intraocular pressure, corneal blood staining, and permanent visual impairment if prompt hemostatic and ophthalmologic management are not provided [10].

Here, we report a three-year-old girl with type 3 VWD who developed traumatic hyphema, an extremely rare presentation, with only one similar pediatric case previously described in the literature [11].

Case Presentation

A 3-year-old girl with known history of Von Willebrand disease (VWD) type 3 presented with history of left eye injury, decreased vision, vomiting, and abdominal pain for two days. The injury was caused by a metal hanger, after which the eye became watery and painful without swelling or bleeding. The following day parents noted that her eye became swollen, red, and more painful with progressive visual loss, so brought her to emergency department (ER). Her past medical history was significant for multiple emergency visits and hospitalization for recurrent epistaxis and gum bleeding that was managed with VWF/F VIII concentrate infusion and tranexamic acid. The parents are cousins, and she has 5 healthy siblings and a paternal cousin diagnosed with VWD type 3.

On arrival at ER, she was vitally stable and looked in distress because of ocular pain. Ophthalmologic examination revealed left upper and lower eyelid swelling with ecchymosis, conjunctival erythema, and subconjunctival hemorrhage (Figure 1 and 2). Extraocular muscle movement was normal in both eyes. Digital palpation of the left eye estimated intraocular pressure (IOP) to be approximately 40 mmHg, compared to 18 mmHg in the right eye. Ophthalmic lens showed a three-quarter hyphema (Figure 3).

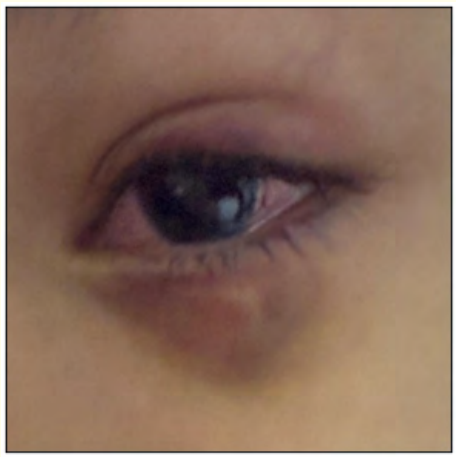

Figure 1

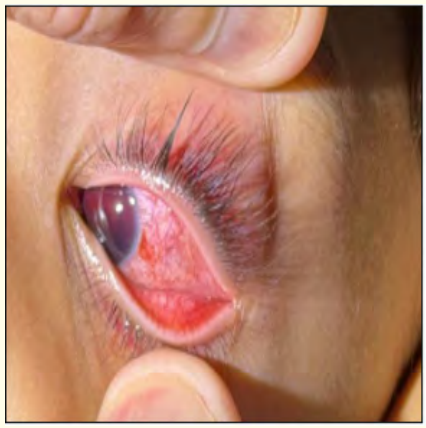

Figure 2

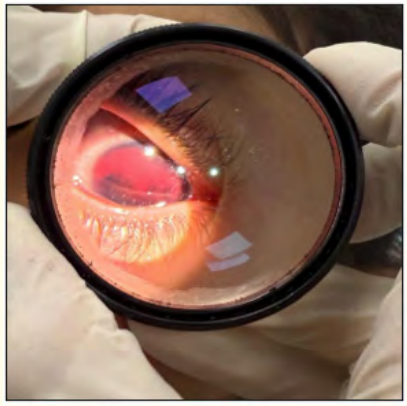

Figure 3

Ocular ultrasonography (US) showed no evidence of vitreous hemorrhage or retinal detachment. CT of the orbit demonstrated minimal left pre-septal oedema without globe rupture or intracranial pathology.

In the ER, she received 500 units of VWF/FVIII concentrate (Wilate), paracetamol, and ondansetron. Ophthalmology recommended strict bed rest, eye shield, a single dose of IV acetazolamide, and initiated topical therapy including cyclopentolate, dexamethasone/tobramycin, and dorzolamide/timolol. The patient was admitted to hospital and received VWF/FVIII (Wilate) every 12h, along with the other supportive management. On day 4, She showed clinical improvement of her eye bleeding, with reduction of hyphema from three-quarter to half, less dense blood collection, and her IOP decreased from 40 mmHg to 20 mmHg, allowing for discharge with a plan of close monitoring and to start prophylactic factor replacement 2-3 times/week. Six days later during her follow-up visit, examination showed rebleeding with total hyphema and new intravitreal hemorrhage. Mother reported difficulty in applying her topical eye drops at home as the child was crying vigorously and rubbing her eyes, however she denied new trauma to the eye. The patient was readmitted to hospital, was managed with VWF/FVIII concentrate 500 units/12 hours (37 u/kg) and tranexamic acid 10 mg/kg/8 hours. Monitoring FVIII level and VWF levels 30 minutes after infusion confirmed good recovery of the factors, ruling out the possibility of inhibitor development (FVIII level > 150%, VWF 97%), after stabilization of her bleeding, VWF/FVIII concentrate infusion was spaced to 24 hourly, with a trough level of VWF 22%, and FVIII 88%.

Repeated ophthalmology follow-up focused on keeping normal IOP, with surgical intervention only if conservative treatment failed. The child was maintained on daily factor replacement for 6 days, then her parents decided to continue management in their home country, for socioeconomic reasons.

Discussion

Although VWD is the most common inherited bleeding disorder, ocular manifestations are rarely reported. To our knowledge, this is the first reported case of traumatic hyphema in a pediatric patient with type 3 VWD, highlighting a unique and clinically challenging management.

Ocular manifestations in pediatric VWD included vitreous, peripapillary retinal and subretinal hemorrhages, reported in only three pediatric cases [12,13]. Traumatic hyphema has been documented once in a 15-year-old girl with mild type 1 VWD (VWF ≈ 35%), who presented with grade III hyphema and elevated IOP following blunt trauma [11]. On the other hand, no adult cases of hyphema associated with VWD have been reported to date, making our case unique in both age and disease severity.

Traumatic hyphema, a significant ocular injury in children, results from disruption of vessels in the iris or ciliary body, leading to blood accumulation in the anterior chamber. While blunt trauma is the most common cause, penetrating injuries and spontaneous cases may occur, especially in the context of underlying conditions such as bleeding disorders (e.g. VWD, hemophilia), sickle cell disease, leukemia, or with anticoagulant use [14].

Our patient presented with symptoms of severe traumatic hyphema; ocular pain, decreased vision, and visible blood in the eye, 2 days after a blunt trauma to her left eye. Her initial examination revealed grade 3 hyphema with significantly elevated IOP of 40 mm Hg. This severe affection is mostly related to her severe bleeding tendency and worsened by the delay in seeking medical attention for factor replacement therapy. Although parents were extensively counseled before on the need to present immediately to ER in case of trauma, they did not appreciate the severity of the condition, being reassured by no obvious external bleeding. They presented late, only after noting that the child had significant decrease in visual acuity in the left eye.

As the child has severe bleeding phenotype, her parents were counseled earlier regarding the need to start her on prophylaxis with VWF/FVIII concentrate infusion 2 - 3 times/week, however due to financial constraints, they could not start prophylaxis and continued On-Demand therapy.

After initial improvement, our patient presented after one week with rebleeding manifested as worsening hyphema and new vitreous hemorrhage, which required admission for further management. This presentation is consistent with the typical period during which early complications arise, especially in patients with known bleeding disorders.

Initial management aimed primarily at preventing complications, as no intervention has demonstrated a clear benefit in improving visual acuity according to current evidence [10]. The patient was treated conservatively with strict activity restriction, head elevation (≥30°), protective eye shielding, and topical agents to reduce inflammation and intraocular pressure. Hemostatic management using tranexamic acid was administered to minimize the risk of secondary hemorrhage, and factor replacement therapy with VWF/ FVIII concentrate started with monitoring of trough and peak levels. As surgical intervention has increased risk of bleeding, it is reserved for patients with uncontrolled IOP despite medical therapy [10]. and in view of her improvement, no surgical intervention was needed initially.

While most cases of traumatic hyphema resolve with conservative care, patients with bleeding tendency are at markedly higher risk of rebleeding, persistent elevation of IOP, corneal blood staining, secondary glaucoma, and potential permanent visual loss. Managing

pediatric patients is very challenging; due to poor cooperation, necessitating careful prioritization of essential tests, cautious examination, and tailored multidisciplinary care. In comparison to most patients who can be managed on an outpatient basis, our patient needed in-patient care due to her severe bleeding disorder and poor compliance.

In pediatric patients with VWD3 presenting with traumatic hyphema, multidisciplinary management is essential to optimize outcomes. Ophthalmologic interventions focus on grading hyphema, monitoring IOP, and managing complications [15]. Major acute complications include intraocular hypertension and rebleeding (within 2-7 days), which may cause corneal blood staining, secondary glaucoma, and optic atrophy. Long-term complications include uveitis, amblyopia, and decreased vision [16]. Worse outcomes are linked with higher grades of hyphema and associated posterior segment injuries [14]. Hematology interventions on the other hand aim at immediate correction of coagulation defect with VWF/FVIII concentrates, adjunctive antifibrinolytics to further reduce bleeding risk, frequent monitoring, including serial assessment of VWF and FVIII levels, screening for inhibitor development, and vigilance for thrombosis risk [6]. Close coordination between hematology and ophthalmology is required for timing and dosing of hemostatic therapy.

As patients with type 3 VWD have no endogenous production of VWF, there is a possibility of inhibitor development with repeated exposure to exogenously infused VWF concentrates. Inhibitor development was reported in around 5 - 10% of patients with type 3VWD complicating their management and rendering treatment ineffective. When our patient presented with rebleeding, the possibility of inhibitor development was thought off. Unfortunately assays for VWF inhibitors are not standardized and are not available in our lab. Nevertheless, this possibility was unlikely in view of the good post infusion recovery and trough levels of factor VIII and VWF and good response to treatment [17].

Although established protocols remain the standard of care, there is limited data on hyphema management in pediatric VWD, and further prospective studies are needed to clarify optimal strategies and long-term outcomes in this rare condition.

Conclusion

This case represents a rare incidence of traumatic hyphema in a child with VWD type 3. It highlights the critical role of early recognition, appropriate replacement therapy, and multidisciplinary coordination to prevent vision-threatening complications. In the absence of specific treatment guidelines for the management of ocular bleeding in VWD type 3, this report raises clinical awareness and supports the need for future research to develop standardized management approaches to improve patient outcomes.

Patient Consent Statement

Informed consent was obtained from the mother to report this case.

Ethics Statement

Ethical approval from Institutional Research Board was obtained to report this case.

Financial Disclosure

Authors have no financial relationships relevant to this article to disclose.

Conflict of Interest

Authors have no conflicts of interest relevant to this article to disclose.

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