Abstract: Traumatic hyphema, or blood in the anterior chamber, is a common complication of blunt injury to the eye and can result in permanent loss of vision. Management of traumatic hyphema in von willebrand disease is a challenging clinical entity encountered by the ophthalmologist. We present the case of an 8 year old boy, known case of von willebrand disease type III, who presented to us with grade II hyphema with raised intraocular pressure in his left eye following blunt trauma with a stick. He responded well to topical steroids, atropine ointment, antiglaucoma medication and oral tranexamic acid. He also received a 4 day course of intermediate purity factor VIII 500IU infusion after which his factor VIII level was restored. But after a day of stopping factor support, rebleed in the form of hyphema and raised IOP was noted. Following this factor support was continued for the next 5 days and his hyphema started resolving with normal IOP over follow up period of 4 weeks. Visual acuity returned to 6/6 with no rebleed into anterior chamber. Extended course of factor support is required in vWD to avoid the risk of rebleed.

Keyword: Von willebrand disease, hyphema, factor VIII

Introduction

Von Willebrand disease (vWD) is one of the most common inherited coagulation disorders, characterized by quantitative or qualitative defect of von Willebrand factor (vWF), a high molecular weight glycoprotein which plays a major role in the early phases of haemostasis by promoting platelet adhesion to the subendothelium and platelet aggregation under high shear conditions [1]. Since vWF is also the carrier of factor VIII (FVIII) in the plasma, deficiency of vWF results in impairment of both the primary phase of haemostasis and blood coagulation. vWD has 3 subtypes: Type 1 vWD refers to partial quantitative deficiency of vWF, type 2 vWD refers to qualitative deficiency of vWF and type 3 vWD refers to virtually complete deficiency of vWF. In many patients with type 1 or type 2 vWD, the bleeding tendency may be absent. In contrast, patients with type 3 vWD have a severe haemorrhagic tendency which may be life threatening. The inheritance pattern of type 3 vWD is autosomal recessive and its prevalence is approximately 1 per million population [2,3,4]. Laboratory findings include reduced activity of vWF, factor VIII or vWF antigen and prolonged bleeding time count with normal platelet count, prothrombin time, activated partial thromboplastin time [5]. In vWD type 1, the treatment is substitution of desmopressin (DDAVP) if vWF activity is higher than 10%. In severe cases, if vWF activity is lower than 10% and in vWD type 2 and type 3, VWF has to be substituted directly [6]. It has been recommended that in the presence of even minor signs of intraocular hemorrhage, the deficient clotting factor (or cryoprecipitate) should be infused regularly during the high-risk period for secondary hemorrhage (i.e., the first 5 to 7 days after the injury). Rebleed occur in 3.5 to 38% of patients in traumatic hyphema during 2nd to 5th day of injury. It is associated with poor visual prognosis. If the patient must undergo surgery to evacuate the hyphema, it may be worthwhile to provide replacement therapy sufficient to restore levels of clotting factor levels to 100% of normal during the procedure [7].

Case Report

An 8 year old boy, known case of von willebrand disease type III, presented to us on the day of injury with a stick, with complaints of painless defective vision in the left eye. He was born to parents who had a 3rd degree consanguinous marriage and who were asymptomatic. His older sister (12 years) and older brother (9 years) were also diagnosed as Von Willebrand disease Type III after an episode of trivial trauma leading to haemarthrosis. On examination, vision in his right eye was 6/6 with normal anterior and posterior segment. In his left eye he was found to have vision of perception of light with accurate projection of rays. Anterior chamber examination revealed grade II hyphema (figure 1). Intraocular pressure recorded was 42mm Hg with no view to posterior segment. B scan showed presence of vitreous haemorrhage in his left eye.

Management:

He showed a good response with topical steroid (Prednisolone acetate), atropine ointment, antiglaucoma medication (IV Mannitol, topical Dorzox T), and oral tranexamic acid. He also received a 4 day course of intermediate purity factor VIII 500 IU infusion till the level of factor VIII was restore to 100% of the
normal level. But after a day of stopping factor support, rebleed in hyphema and raised IOP (28mm Hg) was noted (figure 2). Following this factor support was continued for the next 5 days. During the next 1 week of hospital stay his hyphema started resolving with controlled IOP (Figure 3). He was discharged with a visual acuity of 6/12 and IOP of 19 with clear cornea and hazy view to fundus due to vitreous haemorrhage (Figure 4). In next 4 weeks of follow up visit his vision improved to 6/6 with resolved hyphema and resolved vitreous hemorrhage.

**Discussion:**

vWD is a common inherited bleeding disorder and may complicate the management of trauma patient as trivial injury can lead to life threatening complication. In the management of traumatic hyphema in patients with von willebrand disease, major concern is to prevent rebleed and its associated complications such as increased intraocular pressure, corneal blood staining, optic atrophy, and peripheral anterior synechiae. Tranexamic acid stabilizes the fibrin clot, by inhibiting activating substances in plasma that convert plasminogen to plasmin, thus preventing rebleeding while permanent vessel repair takes place [8]. As seen in our case, factor VIII transfusion for the traditional four days and stopping thereafter may lead to rebleed and raised IOP. This was reversed with an extended course of factor VIII support. While this needs to be researched further, we therefore suggest that factor VIII support to be maintained for 10 days in such situations to tide over the high risk period for rebleeds and thereby prevent sight threatening complications.

**References:**
