

APPROACH TO A CASE OF CORNEAL TEAR IN A PATIENT OF HEMOPHILIA A.

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ABSTRACT

Purpose – To report an unusual case where the diagnosis of haemophilia was made after corneal tear repair and intractable hyphema **Methods**- A 21year gentleman presenting with corneal tear and raised intraocular pressure after trauma was taken up for corneal tear repair but intractable bleeding was noted during surgery. The blood tests done preoperatively were normal. **Results** – During corneal tear repair, intraoperative bleed could not be controlled with air or Ophthalmic viscoelastic device (OVD) tamponade. On post operative day 1, full chamber hyphema was noted. Given the unusual bleeding, the patient was referred to the physician for further evaluation. Laboratory studies revealed a diagnosis of haemophilia A. **Conclusion**- This case report illustrates the difficulties in management of ocular trauma in a patient of Hemophilia A and importance of history taking and management of systemic conditions before planning for surgery.

KEYWORDS: Corneal tear, Hemophilia A.

INTRODUCTION

Ocular trauma can be in form of open globe injury or closed globe injury. The proportion of open versus closed globe injury varies in different studies. However, it is generally accepted that open globe injury results in more hospitalisation and poorer visual outcome than closed globe injury.

Classification of open globe injury

Type (Mechanism of injury)	Grade (Presenting visual acuity)	Pupil (RAPD)	Zone (Location of wound)
Rupture	>20/40	Positive – RAPD present	I – cornea
Penetrating	20/50 to 20/100	Negative – RAPD absent	II – Limbus to 5 mm posterior into sclera
Intraocular foreign body	19/100 to 5/200		III – Posterior to 5 mm from limbus
Perforating	4/200 to light perception		
Combined	No light perception		

A small corneal tear has good visual prognosis but if associated with bleeding disorder it can still be vision threatening.

Hemophilia A is the oldest recognized hereditary bleeding disorder. It is sex-linked in transmission. The gene for hemophilia A is located on the long arm of the X chromosome. Its incidence is 1 in 5,000–10,000 male births. Hemophilia A is classified based on the amount of factor VIII activity. Patients with severe hemophilia A (<1% factor VIII activity) have joint bleeding with resulting hemarthroses as well as deep intramuscular

bleeding. The main presenting feature is bleeding. Bleeding into joints (hemarthrosis) is characteristic. Other sites are muscles, digestive tract and brain. The ocular manifestations can be in form of spontaneous bleed as in subconjunctival hemorrhage, optic disc hemorrhage, retro bulbar hemorrhage or hyphema. The bleeding can also present after trauma in form of hyphema. These complications are often correlated with minor traumas. Because the ocular complications in haemophilia are very rare^{1,2}, it is of importance to present the case of a haemophilic young male with corneal tear and raised intra-ocular pressure after trauma.

CASE REPORT

A 21 year male presented to our clinic with complaints of decreased vision and pain since 1 day after injury to the left eye with a metal wire. On examination the vision in left eye was PL+ PR inaccurate. Slit lamp biomicroscopy showed conjunctival congestion along with radial corneal tear between 4-5 o' clock with iris prolapse at the wound. Anterior chamber had inferior blood clot with fibrin membrane over iris. There was no view of the iris. IOP recorded was 44 mm of Hg. B-scan showed cataractous lens echo (? subluxation) along with low echospike reflective membranes in vitreous cavity and attached retina. We admitted the patient and started on oral and topical medications and planned for corneal tear repair with anterior chamber wash. Blood investigations (CBC, BT, CT) were all within normal limits. However patient gave history of bleeding more than normal after an injury.

Intraoperatively iris was abscised and corneal tear sutured with two interrupted 10-0 nylon sutures. Severe bleeding was there not responding to tamponade with air/OVD. The iris details were not clear (? 270°iridodialysis with attachment inferonasally) and crystalline lens found to be subluxated posteriorly.

On postoperative day – 1, vision was PL+PR inaccurate with IOP of 42 mm of Hg. There was full chamber hyphema. (Fig 1).



Fig. 1: Full chamber hyphema.

On post operative day – 2, there was severe conjunctival chemosis with full blood clot in anterior chamber. IOP was digitally high.(Fig 2).



Fig. 2: Severe conjunctival chemosis with full chamber blood clot.

Physician consultation was sought. He took the history of spontaneous bleeding from nose, gums, into joints since

childhood (Fig 3,4) and he advised for blood tests – CBC, PT/INR, APTT.



Fig. 3 and 4: shows joint deformities.

APTT was raised – 64.6 sec (control – 27.6 sec). Factor VIII and IX assay was advised. Factor VIII was decreased (3.4%) while factor IX (76.4%) was normal. Based on the blood reports a diagnosis of Hemophilia A was made and factor VIII transfusion was advised.

On review after 3 weeks (post factor VIII infusion), vision was PL+PR inaccurate with IOP 30 mm of Hg. There was severe conjunctival chemosis with full chamber blood clot in anterior chamber. Glaucoma consultation was sought and was continued on anti-glaucoma medications. We decided to plan Transscleral cyclophotocoagulation (TSCPC) if IOP does not decrease.

DISCUSSION

The presented case is interesting because the patient presented for the first time after ocular trauma. He was not diagnosed earlier as a case of haemophilia. The intractable bleeding during surgery and also the postoperative full chamber hyphema made us to ponder over the diagnosis of bleeding diathesis. The intraocular pressure was raised pre and post operatively even after maximum possible anti-glaucoma medications because of full chamber hyphema. History taking is very important in these cases as cause of bleeding can be elicited from history.

Haemophilia is a severe and rare haemorrhagic syndrome, with an incidence of 1 in 10,000 cases, and occurs especially in male children. The characteristic clinical aspect in the severe form includes haemorrhages (haematomas) more often intraarticular, seldom

intramuscular, subperiosteal, intracranial or in the soft tissues (subcutaneous fat)^[1,3] The typical symptom is represented by pain and, in case of soft tissues, local swelling in the area affected by the haematomas. The implications of haemophilia in ophthalmology are very rarely mentioned in the literature^[4,5] and represent an unusual cause of haemorrhage/haematoma in this area.

There have been various case reports of ocular presentations post trauma in haemophilia patients. White *et al.*^[4] described a case of orbital haematoma after retrobulbar anaesthesia at a patient with haemophilia A, haematoma that occurred 12 hours after cataract surgery; the case ended with loss of vision at the involved eye, despite the ophthalmological and haematological treatment provided. Chio *et al.*^[6] described a 360 degree, perilimbal subconjunctival haemorrhage, that occurred during a combined keratoplasty and cataract surgery, at a 75-years old patient who didn't present any signs (anamnestic or clinical) of haemophilia. Zolog *et al.*^[9] described a case of an 11-year old boy, suffering from mild haemophilia type A. The patient developed acute severe unilateral exophthalmos after a local minor trauma. Subsequent haematological treatment led to the complete remission of the ocular phenomena within 6 weeks, except for the lowering of the visual acuity caused by the optic nerve involvement due to the orbital haematoma compression. Ocular or any other surgery should always be planned only after factor VIII transfusion. The recent introduction of recombinant factor VIII replacement therapy has immensely improved the management of hemophilia patients.^[10] One complication of replacement therapy, however, continues to present a challenge: the development of factor VIII inhibitors in a large percentage of severe hemophilia A patients.

Severity	F VIII level ¹	F VIII level ²	Clinical presentation
Severe	<2%	<1%	Spontaneous haemarthroses and muscle haematomas
Moderate	2-10%	1-5%	Mild trauma or surgery causes haematomas
Mild	10-50%	5-25%	Major injury or surgery results in excessive bleeding

CONCLUSION

The corneal tear being so small resulted in vision loss as diagnosis of hemophilia could not be made preoperatively in our case. It made us to think about proper history taking and management of systemic conditions before planning for surgery.

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