Case Report

Retinal Detachment Surgery in the presence of Haemophilia under regional anaesthesia

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ABSTRACT

Objective: Patients with hereditary bleeding disorders rarely present with intraocular complications. Conducting posterior segment surgery in a haemophiliac, where achieving a perfect haemostasis is in jeopardy and in turn, poses a challenge to predict the outcome of surgery. Awareness of retinal surgical management of such conditions can improve surgical decisions.

Case description: An 55 year old gentleman, known case of haemophilia and who has recovered from COVID infection recently was posted for retinal detachment surgery in his left eye (LE). At the age of seven, he was diagnosed to have haemophilia with multiple joint haemarthroses and arthropathy. His present co-morbid conditions include Type 2 diabetes and hypertension. As per the protocol drawn by the Haematologist, on the day of surgery, 2,250 IU of the recombinant factor VIII [Eloctate] was administered slowly over fifteen minutes. Thirty minutes later, patient was shifted to operation theatre. Under episcleral block, 25 G vitrectomy was conducted on a phakic myopic eye with inferior retinal detachment and peripheral tear. Retinotomy was performed to remove the band and drain all the fluid. Surgery was completed after tamponading with silicon oil. His post operative recovery of the eye condition was found to be satisfactory.

Conclusion: Bleeding disorders present a dilemma in the surgical management of retinal detachment in a high myopic with a known case of hemophilia and recent Covid exposure. Close follow up with Haematologist to conduct the retinal surgery under the cover of reconstituted recombinant Factor VIII went a long way in achieving the desired results.

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Introduction

Haemophilia is the most common and serious X-linked inherited coagulation factor deficiency. Multiple subtypes of haemophilia exist, 85% of which are of the factor VIII deficiency or haemophilia A type. Clinically, however, haemophilia are all virtually identical and manifest in the same way, and is due to the similarity in their patho-physiologies, affecting the same coagulation pathway of the human body, albeit at different points for each type.[1] A deficiency of a factor in a certain step identifies the type of haemophilia or manifesting as prolonged bleeding into the joints and muscle. Other bleeds, which include ocular bleeding and trauma, comprise only 5% of reported cases of initial presentation of haemophilia. Ocular haemorrhage can be from both traumatic and non-traumatic causes. Diagnosis and management of haemophilia in consultation with the haematologist is more vital in these patients who are to undergo ocular surgery.[2]

In 2014, a meta-analysis[3] found that orbital haemorrhage was attributed to inherent bleeding disorders in only 19% of reported cases in literature over the past 30 years. Of these 24 cases, only 0.8% of the total was due to haemophilia, showing the rarity of non-traumatic ocular bleeding as a presentation of haemophilia. Traumatic orbital haemorrhage is more common. However, bleeding from ocular trauma as the only initial manifestation of an underlying haemophilia condition is rare.[4]

Haemophiliac patients usually present with other forms of prior bleeding episodes. Paying attention to disproportionate decrease in the visual acuity to the amount/manner of trauma should therefore alert the attending ophthalmologist to the presence of a more serious underlying condition. In haemophiliac patients, continuous traumatic orbital bleeding causing proptosis of the globe could actually develop into total peribulbar haemorrhage within 24 hours.[5]

Case Description

A 55 year old business man presented with complaint of diminution of vision in the left eye (LE) due to retinal detachment for 6 months. He had postponed the immediate surgical intervention due to fear of the prevailing Covid pandemic. Subsequently, he had Covid infection and was hospitalised for treatment. His left eye is myopic since childhood; and he was treated for haemophilic multiple joint hemarthroses, arthropathy from the age of 14 years. This resulted in painful and restricted right elbow, left hip and knee joints.

His present co-morbid conditions include type 2 diabetes and hypertension. His surgical history included left knee and hip replacement ten years back. Three years back, he underwent laparotomy to drain left psoas hematoma for peritoneal bleed. Following laparotomy, he developed urinary infection, renal failure, type 1 septicaemia and blood transfusion induced Hepatitis C.

He gave no history of ocular trauma or surgery Family history confirms that his 50 years old cousin is also a haemophiliac. He presented for retinal detachment surgery with treatment protocol for haemophilia before surgery and with arrangements to report back to his haematologist at a Medical centre after surgery for further care and follow-up. His preoperative eye examination revealed: high myopia in both eyes and LE also had sub-total rhegmatogenous retinal detachment with macula off and proliferative vitreo-retinopathy (PVR).

On admission, a detailed informed consent was obtained for vitrectomy to address the detachment in the LE under episcleral block. He was counselled about the indication for the proposed surgery to save his vision; and the unforeseen prognosis in presence of his bleeding disorder.

As per the protocol drawn by the Haematologist, on the day of surgery 2,250 IU reconstituted recombinant factor VIII [Eloctate] was administered slowly over 15 minutes. Thirty minutes later, the patient was shifted to operation theatre. Episcleral block was administered with 4 ml of 2% Lignocaine with 1 in 200,000 adrenaline, 200 IU of hyaluronidase and 4 ml of 0.75% Ropivacaine. 25 G vitrectomy was conducted on a phakic myopic eye with inferior retinal detachment and peripheral tear. Retinotomy was performed to remove the band and drain all the fluid. Surgery was completed after tamponading with silicon oil.

The anaesthesia concerns included the possibility of retrobulbar hemorrhage with serious consequences following needle or cannula based regional anaesthesia block. He was discharged and sent back to his haematologist at the Medical Institute. At the Institute, he received an additional dose of recombinant factor VIII before discharge. The post operative course was found to be uneventful expect for high intraocular pressure on day seven, which was managed medically.

Discussion

Robert A Rubenstein et al[6] reported ocular complications in 123 patients with hemophilia. In one of these patients severe spontaneous retrobulbar hemorrhage resulted in loss of vision in the affected eye. Prolonged bleeding followed extraocular muscle surgery, enucleation, chalazion surgery, and cataract extraction was observed in other patients. In addition, 20 patients with haemophilia had sub-conjunctival haemorrhage or other haemorrhages in the eye.

Jijina F et al[7] published their experience of eye surgery in five patients with haemophilia of varying severity aged between 8 and 75 years. The surgery included intraocular lens implantation, trabaculectomy and vitrectomy. They reported successful outcome with haemostasis with moderate dose of clotting factor concentrate along with oral epsilon aminocaproic acid and IV desmopressin wherever feasible.

They also treated a patient with 32% of factor VIII presenting with hyphaema following ocular surgery conducted without knowing the underlying hemophilic condition. They conclude that satisfactory eye surgery is possible in presence of haemophilia with recombinant factor VIII therapy.

Luis Miquel Aquino and Felice Katrina Ranche[5] reported an eight year old Filipino boy presenting with traumatic hyphaema with corneal staining following a blunt trauma in the right eye. Sub-retinal hemorrhage was seen on ultrasound. The patient underwent anterior chamber washout with temporary keratoprosthesis and pars plana vitrectomy with silicon oil tamponade. Nineteen days later, he presented with recurrence of hyphema with a new onset of proptosis and retrobulbar hemorrhage. A bleeding disorder was suspected at this point. Further probing revealed a family history of prolonged bleeding time in an X-linked genetic inheritance pattern spanning three generations. Laboratory testing revealed factor VIII deficiency, diagnostic of haemophilia A. No further surgery was done.

Our 55 years old wheelchair bound haemophilic patient presented with complaint of diminution of vision in the LE for six months due to retinal detachment and also had type 2 diabetes and controlled hypertension. He was treated for multiple joint haemarthroses from the age of 14 and had undergone left knee and hip replacement and laparotomy for left psoas hematoma. Meanwhile he had Hepatitis C, urinary infection, renal failure and type 1 septicaemia. Recently he suffered Covid infection. He gave no history of ocular trauma or surgery. He presented for RD surgery with haemophilia treatment protocol from his referring haematologist. After surgery, he was admitted at medical institute for further care and follow-up.

Patient received recombinant factor VIII 30 minutes before shifting to OT. Our patient did not have petechial lesion or haemorrhagic discoloration at the IV cannulation site indicating presence of near normal clotting process. 25 G Micro-Incision Vitrectomy Surgery (MIVS) was conducted on a phakic myopic eye with inferior retinal detachment with peripheral tear under episcleral block. We planned to modified ocular surgery in this patient with haemophilia like regular circumferential scleral buckling procedure as part of retinal surgery was not considered since it involved extensive dissection and handling of structures in presence of haemophilia; routine pneumatic retinopexy with air or heavy gas tamponading was not considered since the patient had to remain in face down position for a long period of time. We performed retinotomy to remove the band and drain all the fluid. Surgery was completed after tamponading with silicon oil. After surgery, he received an additional dose of recombinant factor VIII before discharge.

His post operative course was found to be satisfactory, except for high intraocular pressure on day seven, which could be managed medically.

Conclusion

Bleeding disorders like haemophilia present a dilemma in the surgical management of retinal detachment in a high myopic. Our patient presented with a history of recent Covid infection with implication of cytokine storm and haemorrhages in the lungs. Close liaison with haematologist and following a well drawn protocol to conduct the retinal surgery under the cover of recombinant Factor VIII goes a long way in achieving the desired results.

References

1. World Federation of Hemophilia. Protocols for the Treatment of Hemophilia and Von Willebrand Disease. 3rd Edition. Georgia: World Federation of Hemophilia;2008.
2. McNab AA. Nontraumatic orbital hemorrhage. Surv Ophthalmol 2014 Mar-Apr;59(2):166- 84.
3. Brooks AM, Finkelstein E. Spontaneous Orbital Hemorrhage. Br J Ophthalmol 1984 Nov;68(11):838-40.
4. French C, Eperjesi F. Ocular complications of haemophilia. Optometry Today 2013 Nov1:55-9.
5. Aquino LM, Ranche FK. Hemophilia presenting as recurrent ocular hemorrhage. GMS Ophthalmol Cases 2020;10:Doc15.
6. Eyster M, Gill FM, Blatt PM. Central nervous system bleeding in hemophiliacs. Blood 1978;51:1182.
7. Robert A Rubenstein; Danial M Albert, and Herold Scheie : Ocular Complications of Hemophilia : Arch Ophthalmol 1966;76;(2):230-2.