Retrobulbar haemorrhage following blunt trauma in a newly diagnosed haemophilia patient: Case report

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ABSTRACT

Haemophilia is a congenital disorder of coagulation. It presents with numerous and varied systemic manifestations depending on the severity of coagulation factor deficiency. There are also ocular manifestations ranging from simple subconjunctival haemorrhage to more debilitating complications like retinal, vitreous and retrobulbar haemorrhage which are potentially blinding.

We report a patient who initially presented with what appeared to be simple post-traumatic retrobulbar haemorrhage. The diagnosis of a bleeding disorder was considered when difficulties in achieving haemostasis were encountered intra-operatively.

Key words: **Retrobulbar haemorrhage, haemophilia, trauma, proptosis**

CASE REPORT

A 5 year old boy from Taita Taveta, Kenya, presented to Kenyatta National Hospital on 28th April 2011 with post traumatic swelling of the left eye for three weeks. He was hit by a stone flung with a sling to his left eye three weeks before presentation and was taken to the local hospital immediately and later taken to Coast Provincial General Hospital (CPGH) for further management.

At CPGH, a diagnosis of left orbital haemorrhage was made. A plain oribital X-ray was done and it showed a linear fracture of the orbital floor. Conservative management of the haemorrhage was recommended. There was no improvement of the orbital swelling and the patient was taken to theatre 5 days later for decompression. There was excessive bleeding intra-operatively and had to be transfused two units of blood.

On further inquiry, it was established that the boy had experienced several episodes of spontaneous calf and arm swelling but no history of excessive bleeding from minor injuries. A coagulation profile was ordered and the results are as presented in Table 1.

With a raised aPTT pointing towards a possibility of haemophilia, the patient was referred to Kenyatta National Hospital (KNH) for further management. On examination at presentation in KNH, the visual acuity in the right eye was 3/3 (Lea's chart) and the left eye had no perception of light (NPL). The right eye had a normal anterior and posterior segments.

The lids of the left eye had a scar over the left upper lid (from surgical incision for orbital decompression), ecchymosis and matted lashes. The conjunctiva had a purulent discharge, marked chemosis and injection. The cornea had a central ulcer measuring 3.5mm by 4.8mm, associated corneal opacification and surrounding infiltrates. Other details were inaccessible. A diagnosis of left eye traumatic retrobulbar haemorrhage with corneal ulcer secondary to exposure keratopathy in a child with bleeding disorder was made. The patient was admitted to the eye ward immediately and started on hourly ciprofloxacin eye drops, tetracycline eye ointment twice daily and oral augmentin (amoxicillin + clavulanic acid combination).

In consultation with a haematologist, further coagulation profiles and haematological tests were ordered as shown in Table 2.

Table 1: Coagulation profile at CPGH

Test	Results	Normal range
(Activated partial thromboplastin time) aPTT	180 sec	26.2-34.6sec
Prothrombin time/ INR*	18sec / 1.3	13sec / 0.8 – 1.2
Bleeding time	2 min	≤4min
Clotting time	28min	≤11min
Platelet count	465,000 /µl	150,000-450,000/ µl

*INR: International Normalized Ratio

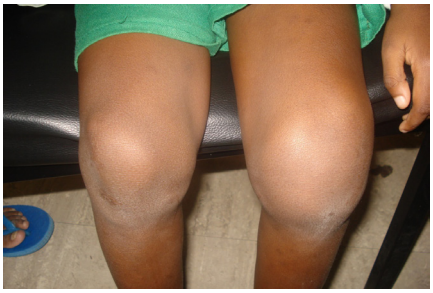
Table 2: Subsequent investigations

Test	Done at CPGH	30/4/11	5/5/11	10/5/11
Prothrombin Time (sec)	18	16	18.4	14.4
PTI		88%	72.8%	91.7%
PTI/INR	1.3	1.4		1.09
aPTT	180 sec (ctrl 26.2-34.6s)	>120sec (ctrl 30s)	222 sec (30s)	74.4sec (34s)
Platelets	Platelets: 465,000	342,000		
Clotting time	28min (≤11min)			
Bleeding time	2 min (≤4min)			
FVIII			3.5% (50-50%)	

From the above tests, it was established that the main derangements were in the aPTT which points to a potential factor deficiency and reduced factor VIII levels, confirming a diagnosis haemophilia A. Because there was no worsening of the haemorrhage, the patient continued with the local therapy with resolution of the corneal ulcer and orbital swelling. The eye became physical.

Three weeks after admission, the patient developed swellings of left knee and left elbow with no history of trauma (Figure 1).

Figure 1: Left knee swelling



There was also associated pain on movement of affected limbs. In consultation with the haematologist, the patient was then started on transexamic acid 500mg eight hourly. A decision was also made to administer Factor VIII if there was increased swelling after 24 hours of monitoring. There was however no further limb swelling noted and the patient was discharged through the haematology clinic and added to the haemophilia registry.

DISCUSSION

Haemophilia is a congenital disorder of coagulation. The most common form of haemophilia is haemophilia A in which there is a deficiency of coagulation Factor VIII. It affects 1:10,000 individuals. The Factor VIII gene is on the X chromosome and inheritance is therefore sex-linked. Other forms of haemophilia include haemophilia B (Christmas disease) in which there is a reduction of Factor IX and haemophilia C (Rosenthal syndrome) in which there is a reduction of Factor XI[1].

Various coagulation profiles are done to establish diagnosis in a patient with suspected bleeding disorder. Of these, aPTT is most specific for haemophilia as it tests for the factors of the intrinsic coagulation system. These are Factors I, II, V, VIII, IX, X, XI and XII. It would also be raised in heparin use, presence of antiphospholipid antibodies and sepsis.

Prothrombin time/INR assesses the extrinsic coagulation pathway and will be raised with the use of warfarin for anticoagulation. Bleeding time assesses platelet function mainly and will be raised in thrombocytopenia, disseminated intravascular coagulopathy and aspirin use, among others. Clotting time assesses mainly for the level or activity of prothrombin and will be raised if prothrombin level or activity is low.

Severity of haemophilia is classified according to the factor activity. According to the classification our patient therefore had moderate Factor VIII activity as his was at 3.5% (Table 3).

Table 3: Factor activity and severity of haemophilia

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

Common systemic presentations: Patients rarely present before 6 months of age because of the relative inactivity of the child before this age. Presentation is related to the factor activity and patients will commonly present with bruising or features of Non-Accidental Injury (NAI).

They also present with haemarthroses. If these are recurrent it results in synovial hypertrophy, destruction of cartilage and secondary osteoarthrosis. Muscle haematomas of the calf and psoas muscle are also common in severe forms of the condition and may eventually lead to muscle ischaemia, necrosis, fibrosis and eventual contracture. Intracranial haemorrhage has also been seen in severe cases[1].

Common ocular manifestation: A case report was made of a child who suffered minor orbito-palpebral trauma with resultant exophthalmos. Complete remission of the exophthalmos occurred but there was loss of vision with optic disc changes, probably due to severe compression by the haematoma on the intraorbital segment of the optic nerve[3]. This is similar in some ways to our case due to the history of trauma with retrobulbar haemorrhage. The trauma in our case however was also not minor because of the associated orbital fracture. This could have led to delayed diagnosis because in most cases an index of suspicion is aroused where the trauma is not expected to cause massive orbital haemorrhage.

Other ocular complications have also been noted in the literature. A case series of 123 haemophilic patients with ocular symptoms showed that some of the more common presentations included severe spontaneous retrobulbar haemorrhage, prolonged bleeding following extraocular muscle surgery, enucleation, chalazion surgery, and cataract extraction[4]. In the same case series 20 patients with haemophilia were noted to have subconjunctival haemorrhage or other haemorrhages about the eye. Ocular and periocular haemorrhages especially after trauma or surgery were also noted. Neuro-ophthalmic signs secondary to central nervous system haemorrhage included pupillary abnormalities, cranial nerve palsies, visual blurring, and papilloedema. Repeated retinal haemorrhages and vitreous haemorrhage was also documented.

Cases of hyphema have also been documented. One was a case of spontaneous hyphema in a child with haemophilia[5] and another a case of recurrent re-bleeds in a patient with hyphema[6]. What appears to be more common are cases of haemorrhage following surgery. These include a case of severe orbital haemorrhage following cataract surgery with total loss of vision[7] and another of a patient who suffered excessive subconjunctival haemorrhage during attempted penetrating keratoplasty[8].

We present this paper as a unique case of a child presenting with the sequelae of haemophilia. It presents a useful learning point in the management of these patients. The sequence of ocular trauma resulting in retrobulbar haemorrhage, worsening on attempted evacuation and eventual corneal ulceration with loss of vision has not been reported in the literature we searched.

In our case we find that no history of bruising or bleeding tendencies was given at presentation. The guardians only gave a history of limb swelling much later and after significant questioning on past medical history. The lack of this history at the initial assessment resulted in the patient being taken for orbital decompression surgery which could either have been avoided or done after adequate preparation with factor VIII and tranexamic acid.

Exposure keratopathy with eventual corneal ulceration and scarring could have been prevented with adequate lubrication and we must therefore keep this very serious complication in mind anytime we are dealing with patients with orbital masses, haemorrhages or lagophthalmos from whatever cause.

Suggested therapies that could have been attempted pre-operatively after considering the level of factor VIII activity and whether the surgery was minor or major with more potential for serious haemorrhage[1]. These include desmopressin, which is a synthetic arginine vasopressin (antidiuretic hormone) analogue. Other than its antidiuretic effect it also increases plasma Factor VIII and von Willebrand factor levels hence its use in haemophilia. Aminocaproic acid and tranexamic acid are both synthetic agents that are orally active and inhibit plasminogen activation, thereby inhibiting the digestion of fibrin allowing for clot formation (Table 4).

Table 4: Pre-operative therapies in haemophilia

Disease severity	Prophylactic treatment
Mild disease (FVIII > 10%).	Desmopressin (IV or Intranasal)
Mild – Severe disease (FVIII < 10%)	
• Minor surgery	Single F VIII infusion + Tranexamic acid for 10 days
• Major surgery	F VIII twice daily for 14 days

CONCLUSION

A high index of suspicion is therefore necessary when dealing with cases of orbital or ocular haemorrhage and a good history and careful examination are really the gems in diagnosing clinical conditions potentially related to haemophilia.

REFERENCES

1. Haslett C, Chilvers E, Boon N, *et al.* Davidson's Principles and Practice of Medicine. 19th Edition. Churchill Livingstone. 2002: 949 – 952.
2. Kaiser RS, Williams GA. Ocular manifestations of hematologic diseases. In: Duane's Ophthalmology [CD-ROM]. Philadelphia: Lippincott, Williams & Wilkins; 2006: Chapter 23.

3. Zolog I, Munteanu M, Gamaniuc E. Particular ocular manifestations in a case of haemophilia Type A. *Timisoara Med J.* [Online Journal] 2003; 2.
4. Rubenstein R, Albert D, Scheie H. Ocular Complications of Haemophilia. *Arch Ophthalmol*. 1966; **76** (2): 230-232.
5. Ghose S, Kishore K, Patil N. *et al*. Spontaneous hyphema in a child with Haemophilia B. *Can J. Ophthalmol.* 1993:**28**: 40-42.
6. Wilker S, Annapurna S, Ellis F. Recurrent bleeding following traumatic hyphema due to mild haemophilia B (Christmas disease) with recurrent re-bleeds. *JAAPOS*. 2007; **11**: 622-623.
7. White W, Mundis R. Delayed orbital haemorrhage after cataract surgery in a patient with an acquired factor VIII inhibitor. *Am J. Ophthalmol* 2001; **132**:785-786.
8. Choi D, Goldstein M, Driebe W. Diagnosis of haemophilia made after intraoperative bleeding during attempted penetrating keratoplasty in an elderly patient. CLAO J. 2001; **27**(1): 53-54.