Challenges in Perioperative management of spontaneous acute subdural hematoma in haemophilia patients

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Abstract

Intracranial hemorrhage in patients with hemophilia is associated with high mortality and morbidity. We report a case of 15 years old boy with haemophilia A, who presented with a spontaneous acute subdural hematoma and underwent craniotomy for clot evacuation. The patient also received Factor VIII infusions peri-operatively along with other measures, to decrease blood loss.

The patient presented with signs of raised intracranial pressure and received mannitol intra-operatively and postoperatively to prevent brain edema. Hypertonic saline (3 ml/kg of 3% solution) was also given over 30 minutes for brain relaxation. Recommendations for peri-operative preparation and management of haemophilia, especially in the setting of emergency major surgery were also reviewed.

Key words: Anaesthesia, Factor VIII, Haemophilia, Intracranial hemorrhage

Introduction

Intracranial hemorrhage (ICH) is a life threatening complication of haemophilia. The site of bleeding is almost equally distributed among subdural hematoma, intracerebral and subarachnoid hemorrhage.[1] Acute subdural hematoma (SDH) is one of the most lethal form of intracranial insult. Prompt surgical evacuation, when indicated, has better prognosis.[2]

Case Report

A fifteen years old boy weighing 48 kg, presented with sudden loss of consciousness for last 12 hours. The patient was a known case of haemophilia-A, having previous history of spontaneous bleeding into joints, and had received Factor VIII twice in the past. He was otherwise normal. There was no history of trauma or drug abuse or any other surgery in the past.

The patient's heart rate was 56/min and blood pressure 138/74 mmHg. Glasgow Coma Scale (GCS) score was E1M5V1 and pupils were bilaterally mid-dilated, not reacting to light. Patient hemoglobin was 11.2 gm/dl, INR 1.63 and APTT 154 sec. Non-contrast computerized tomography (CT) scan showed an acute left Fronto Temporo-Parietal Sub-Dural Hematoma, with mass effect and a midline shift of 10 mm. (Figure 1)

After shifting to the neurosurgery intensive care unit, patient was intubated with IV fentanyl 100 mcg, propofol 80 mg, vecuronium 4 mg and lignocaine 60 mg and kept on mechanical ventilator. Neuroprotective measures in the form of manitol (1 gm/kg, IV, TDS), seizure prophylaxis (phenytoin 100 mg, IV, TDS) and hyperventilation (to a PaCO2 of < 32 mmHg) were initiated. The patient was posted for an emergency craniotomy and clot evacuation after stabilizing his coagulopathy and hemodynamic parameters. Hematology consultation was also taken and 10 vials (2500 units) of dried factor VIII concentrate were transfused. His APTT was 27 seconds two hours later and then he was taken for surgery.

Inside the operating room, his left radial artery was cannulated. Internal jugular vein was cannulated with triple lumen catheter for central venous pressure monitoring and in anticipation of major blood loss. Monitoring included pulse-oximetry, electrocardiogram, end-tidal carbon dioxide, arterial blood gas monitoring,

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temperature monitoring and urine output. Unilateral scalp block was given by using lignocaine-adrenaline preparation. Anesthesia was maintained with oxygen, air and isoflurane. The patient received fentanyl 100 mcg IV at the start of the surgery, total dose 4 mcg/kg) for analgesia and vecuronium for muscle relaxation. 150 ml (3 ml/kg) of 3% saline was given over 30 minutes for brain relaxation at scalp incision.

The patient underwent left fronto temporo parietal craniotomy and hematoma evacuation.

The patient received 1 vial (250 units) of factor VIII concentrate during surgery. Tranexamic acid (500mg) was also given. Blood loss was about 150 ml. One unit of PRBC was transfused. Serial ABG analysis showed no major acid-base imbalance. The patient remained hemodynamically stable throughout the surgery and was shifted back to ICU for postoperative elective ventilation. Post-operative analgesia was provided with IV fentanyl and Paracetamol infusion.

One vial of factor VIII concentrate was given 2 hourly post operatively on the 1st postoperative day and 3 hourly from the 2nd day of surgery. Patient showed signs of neurological improvement on the 2nd day, with GCS improving to $E_3M_6V_T$, and was weaned off the ventilator on the 3rd postoperative day. Factor VIII levels were measured and reported to be 66% on the 5th postoperative day and 48% on the 14th day. factor VIII concentrate was continued until 14th post operative day. On 21st postoperative day the factor VIII level was 39% . There was no postoperative hemorrhage. The GCS improved to $E_4M_6V_5$ by the 21st day and patient was discharged. Regular follow up was done for 12 months and there was sustained improvement in neurological status of patient.

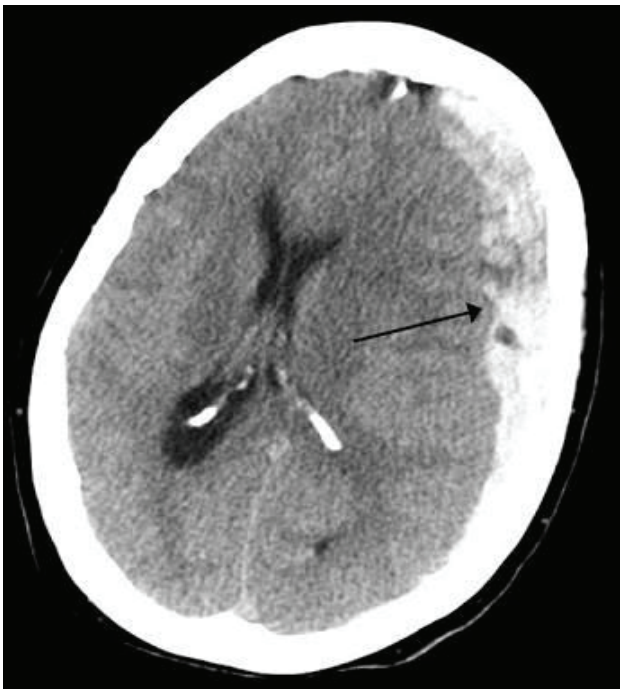

Figure 1: NCCT Brain suggestive of Acute Left frontoparietal subdural hematoma with mass effect and midline shift

Reference author/year	Age M/F	CT/MR	Surgery / Conservative	Peri operative management	Outcome
Agrawal D, 2003 [12]	30y/M	A large acute subdural hematoma in the right parieto-occipital region with gross midline shift and effacement of cisterns	Craniotomy and evacuation	Cryoprecipitate, Freshly frozen plasma, Supportive management	Post operative EDH, EDH evacuation, Good recovery
Meguro T 2004 [13]	54y/ M	Acute SDH with mass effect and mid line shift	Craniotomy and evacuation	Mild haemophillia, Supportive management	Recurrence of SDH, Re exploration, Mild memory loss, homonymous hemianopia
Gyanesh P, 2013 [14]	50 y /M	an acute left temporoparietal sub-dural hematoma (SDH) with left frontal hematoma, with mass effect and a midline shift of 10 mm.	Craniotomy and evacuation	Factor VIII concentrate transfusion, Supportive management	Tracheostomy, Right hemiplegia
Present Study,	15y/M	Acute left Fronto Temporo-Parietal Sub-Dural Hematoma 20 mm thick, with mass effect and a midline shift of 10 mm	Craniotomy and hematoma evacuation	Factor VIII concentrate transfusion, Supportive management	Normal neurological examination

Table 1: Table showing a comparative review of management and outcomes of similar cases in literature

Discussion

Subdural hematoma occurs not only in patients with severe head injury but also in patients with less severe head injuries, particularly those who are elderly or who are receiving anticoagulants. Subdural hematoma can also be spontaneous or caused by a procedure, such as a lumbar puncture. Rates of mortality and morbidity can be high, even with the best medical and neurosurgical care. Subdural hematomas are usually characterized on the basis of their size and location and the amount of time elapsed since the inciting event, age (i.e., whether they are acute, subacute, or chronic). When the inciting event is unknown, the appearance of the hematoma on neuroimaging studies can help to determine the chronology of haematoma (acute/subacute/chronic). These factors, as well as the neurologic and medical condition of the patient, determine the course of treatment and may also influence the outcome. The usual mechanism that produces an acute subdural hematoma is a high-speed impact to the skull. This causes brain tissue to accelerate or decelerate relative to the fixed dural structures, tearing bridging vessels. In elderly persons, the bridging veins may already be stretched because of brain atrophy (shrinkage that occurs with age). Much less common causes of subdural hematoma involve coagulopathies like haemophilia and ruptured intracranial aneurysms .

Hemophilia A, a recessive X-linked disorder involving lack of functional clotting factor VIII (FVIII), represents 80% of Haemophilia cases. Severe cases (<2% of Factor VIII levels) have spontaneous bleeding, predominantly in joints and muscles.

Patients have high APTT but platelet count, bleeding time and prothrombin time may be normal. Factor VIII assay is diagnostic for Hemophilia A as a cause of Intracranial hemorrhage, with incidence of 3-12%,[3] and accounts for over 30% of deaths in Hemophiliacs.[4]

Each FVIII unit per kilogram of body weight raises the plasma FVIII level by approximately 2%, with the half life of 8-12 hours. FVIII should be infused by slow IV, at a rate < 3 ml/minute in adults and 100 units per minute in young children. Cryoprecipitate are the next choice and it contains about 80 units FVIII in volume of 30-40 ml. Fresh frozen plasma may also be used if factor concentrates are unavailable. One ml of FFP contains 1 unit of factor activity.

Tranexamic acid is an antifibrinolytic agent that promotes clot stability and is useful as adjunctive therapy in Hemophilia.

The patient was taken up for surgery at the earliest, after APTT was normalized. We used 3% saline to decrease the cerebral edema as hypertonic saline provides better brain relaxation than 20% mannitol.[5,6] Recent studies also suggest that use of 3% saline in the neurosurgical patients has lesser side effects on the clotting factors and platelets than the use of mannitol.[7] Central venous access, if required, should be secured under ultrasound guidance.[8] We inserted both arterial and central line for better monitoring and patient safety.

Tracheal intubation and airway manipulation in these patients can lead to life threatening submucosal haemorrhages.[9] Early tracheostomy has been found to provide significant benefits in critically ill neurosurgical patients. It reduces the ICU stay, duration of ventilatory support and antibiotic dose requirement in these patients.[10] Extremities and pressure points should be padded to prevent intramuscular hematomas and haemarthrosis. Non-steroidal analgesics were not used. Temperature was monitored and hypothermia was avoided. Controlled hypotension techniques prevent haemostasis of small vessels but are not recommended.

FVIII concentrates were continued in the intra-operative and postoperative period. In Haemophilia, major surgery should take place in a center with adequate laboratory support for monitoring of clotting factor level and preoperative assessment should include inhibitor screening. As our patient needed emergency surgery, FVIII assay and inhibitor screening were not feasible preoperatively. Postoperative monitoring for bleeding was done with hemoglobin and APTT levels along with FVIII assay.

At times despite FVIII concentrate infusion, aPTT fails to improve where we need to think about presence of FVIII inhibitors where plasmapheresis, immunosuppression and steroids are indicated for optimization. [11]

A comparative review with finding, management and outcomes of similar cases in literature has been tabulated as below in table 1.

Conclusion

To conclude, Spontaneous acute SDH may have a number of causes with haemophilia one of the rare causes. A good review of clinical history may provide clue toward underlying cause. To successfully manage such cases, we need Factor VIII levels done in pre-operative period as per feasibility and transfusion of Factor VIII concentrate along with other standard protocols of management along with timely surgical intervention if indicated.

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