Vol 1 No. 2 October 2021
Solo Journal of Anesthesi, Pain and Critical Care

E-ISSN : 2797-0035
P- ISSN : 2776-1770

CASE REPORT

Perioperative Management of Subdural Hemorrhage Trepanation Decompression with Hemophilia A

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Article Info :
Submitted :
26-08-2021
Accepted :
17-10-2021
Published :
30-10-2021
https://dx.doi.org/10.20961/soja.v1i2.54642

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ABSTRACT

Hemophilia is a serious inherited blood disease, transmitted by women, that affects mainly men and lasts for a lifetime. Hemophilia A is the most common form. If any of the factors necessary for blood clotting are absent or insufficient, the clotting mechanism is disturbed, causing insatiable bleeding. The most common cause of death in hemophilia patients is cerebral hemorrhage due to head trauma. In cases of intracranial hemorrhage, surgery should be performed immediately to obtain a better prognosis. A 17-year-old man diagnosed with a 2x4 loss of consciousness due to intracranial subdural hemorrhage (SDH) in the left frontotemporoparietal region and cerebral edema on day 4 accompanied by subfalcine herniation to the right with hemophilia A, planned trepanation decompression for SDH evacuation. The patient received 4000 units of factor VIII injection before surgery. Bleeding during surgery was 1100cc and he received a transfusion of 1940cc blood products until hemodynamically stable. In the postoperative phase, he was admitted to the ICU for 8 days, extubation was performed after the condition improved. In patient with hemophilia, evacuation of bleeding should be performed immediately, but there is a high risk of rebleeding. A recombinant factor VIII substitute should be administered immediately for the treatment of acute bleeding in patients with severe haemophilia A. Anesthetic maintenance should include reducing the risk of hypertension and tachycardia to minimize bleeding.

Keyword: brain injury; coagulation; hemophilia A; subdural haemorrhage

INTRODUCTION

Hemophilia is a serious inherited blood disease, carried by women, acquired mainly in men, and lasts a lifetime. Hemophilia affects people of all races in the world. Compared with hemophilia A, hemophilia B (factor IX deficiency) is less common, occurring every 30,000 male births[1]. Bleeding will easily stop in healthy people, but the formation of clots to stop bleeding is delayed in hemophiliacs. If any of the factors necessary for blood clotting are absent or insufficient, the clotting mechanism is disturbed, resulting in insatiable bleeding. Currently, the most common cause of death among hemophiliac patients is cerebral hemorrhage due to head trauma[1].

Spontaneous intracerebral bleeding is a rare and difficult condition to treat, especially in hemophiliacs. The risk-benefit ratio resulting from the balance between conservative and surgical treatment is low. Surgical treatment carries the risk of rebleeding, while conservative treatment carries the risk of progressive damage and has a high treatment value[2]. Health workers such as doctors and others need to understand hemophilia, blood-clotting mechanisms, and the different types of intracranial bleeding to improve the initial management of patients. Severe hemophilia often manifests clinically in the first few months of life. Whereas mild or moderate hemophilia occurs later in childhood or adolescence, either by accident or after trauma. Hemophilia can manifest as spontaneous intracranial bleeding in the newborn, excessive postoperative bleeding after circumcision, atraumatic painful hemarthrosis, unexplained bruising when the infant crawls or walks, and excessive musculocutaneous bleeding, either spontaneously or after intramuscular vaccination. Intracranial hemorrhage is the most immediate life-threatening manifestation of hemophilia A with the potential for chronic neurological disability and long-term neurological sequelae[3].

Administration of a recombinant factor VIII substitute for the treatment of acute bleeding in patients with severe haemophilia A should be started immediately prior to completing the patient evaluation. Factor VIII replacement calculations for bleeding in severe hemophilia A are: for severe and life-threatening bleeding, administer factor VIII to achieve 100% of the

targeted level; for mild to moderate bleeding, administer factor VIII to achieve a targeted level of 30% to 50%. Factor VIII, if known, should be considered in the initial calculation of factor VIII levels in patients with hemophilia A. Other adjunctive pharmaceutical therapies for bleeding induced by hemophilia A include desmopressin, tranexamic acid, epsilon-aminocaproic acid and factor VIII inhibitors. Intravenous, subcutaneous, or intranasal desmopressin (DDAVP) is useful for treating bleeding in patients with mild to moderate hemophilia A by inducing the release of the von Willebrand factor complex and factor VIII from vascular endothelial cells[3].

CASE ILLUSTRATION

A-17-year old man with a diagnosis of decreased consciousness of SDH with hemophilia A was planned for SDH evacuation decompression trepanation. The patient had a traffic accident and did not lose consciousness 12 hours before admission to the hospital, there was loss of consciousness and difficulty speaking for the next 6 hours, and then the patient was admitted to the hospital for further treatment. Intubation was conduct in the P1 ER due to decreased consciousness and treatment in the Intensive Care Unit in Covid-19 and Tropic Infection Departement for 2 days, the PCR swab screening was negative. Subjective examination showed no allergies and routine medication. The patient had a history of hemophilia A.

Physical examination revealed a patent airway on ETT with breath on ventilator mode PSIMV Peep 3, Pins 8, Psupp 6, FiO2 50% with output SpO2 99% VT 250-320, symmetrical chest wall motion on both sides, pulmonary vesicular sound +/+, rhonchi - /-. Warm, dry, and red tip with CRT <2 seconds, blood pressure measured 135/75 mmHg with pulse 92 beats/minute, 1-2 regular heart sounds and no murmurs or gallops. GCS was still using midazolam sedation 3mg/hour, light reflex +/+ and pupil isocor 3mm/3mm. 550cc urine output on the catheter in 5 hours. The abdomen found neither distended nor tender. Neither edema nor cyanosis of the extremities was found.

The patient was assessed with ASA physical status 4E. Head CT scan revealed subdural hemorrhage in the left fronto temporo occipital region, anteroposterior cerebral interhemisphere, left cerebral tentorium with a thickness of 30mm, 9 slices suspected to be active and

cerebral edema with subfalcine herniation 20mm to the right and transtentorial herniation downward at the level of the mesencephalon and left subgaleal temporal parieloccipital hematoma (Picture 1). From a chest x-ray examination, the heart and lungs were found to be normal (Figure 2). Laboratory results are in Table 1. The patient was planned for trepanation, evacuation, and decompression SDH with general anesthesia and postoperative planning for ICU care. Prior to surgery, the patient received 4000 units of factor VII injection in 40 minutes. Tranexamine acid given during the surgery. The patient was under general anesthesia management and brain protection intubation with controlled ventilation. The patient was positioned head up 30 degrees previously and non-invasive monitors were installed such as blood pressure, heart rate, ECG, and oxygen saturation The drugs used during induction include fentanyl 100 mcg, lidocaine 80 mg, and vecuronium 8 mg. Intubation using ETT no 7.5 cuffed, full control breathing with a ventilator. Analgesics used during surgery were fentanyl syringe 1mcg/kg BW/hour, continuous vecuronium 1-2mcg/kg BW/hour, and sevoflurane as a maintenance agent. The procedure lasted for 5 hours, hemodynamically stable with bleeding of 1100cc. For maintain patient haemodinamic, he received kritaloid 1000cc and colloid 300cc, also blood product transfusion with a total of 1942cc (6 packed red cells and 4 fresh frozen plasma flasks). Postoperative care using ventilator control in the ICU. In the ICU, patient breathing controlled by ventilator PSIMV mode with P Inspiration 10 and P Support 10, PEEP 3, rate 14 times per minutes and FiO2 40%. Tidal Volume and oxygen saturation has been achieved with this setting but initially there was no improvement in patient consiousness. The patient consiousness in 1x2 in GCS. This condition exacerbated by pneumonia so it requires antibiotic treatment according to bacterial culture with Tigecycline 50mg twice daily and Paracetamol 3x1gr for patient fever. After 4 day, we wean the patient from ventilator with PS Spontan mode with P Support , PEEP 3, rate 14 times per minutes and FiO2 40%. Tidal Volume 350-450 and oxygen saturation has been achieved 100% but no improvement in consciousness. We planned for percutaneus dilatational thracheostomy in 10^{th} day but at 7^{th} day the GCS arise from 1x2 to 2x4 and at 8^{th} day we extubate the

patient. During ICU period the patient still get Factor VIII 1000unit in 4 A.M and 1000unit in 4 P.M.

Table 1. Preoperative laboratory

Hb : 10	**Na : 143**
Leukocytes : 3020	K : 4.21
Hct : 30.7%	Cl : 105
Platelets : 206.000	Urea : 41.9
PT 11.3	Creatinine : 0.71
APTT 34.4	Albumin 3.73

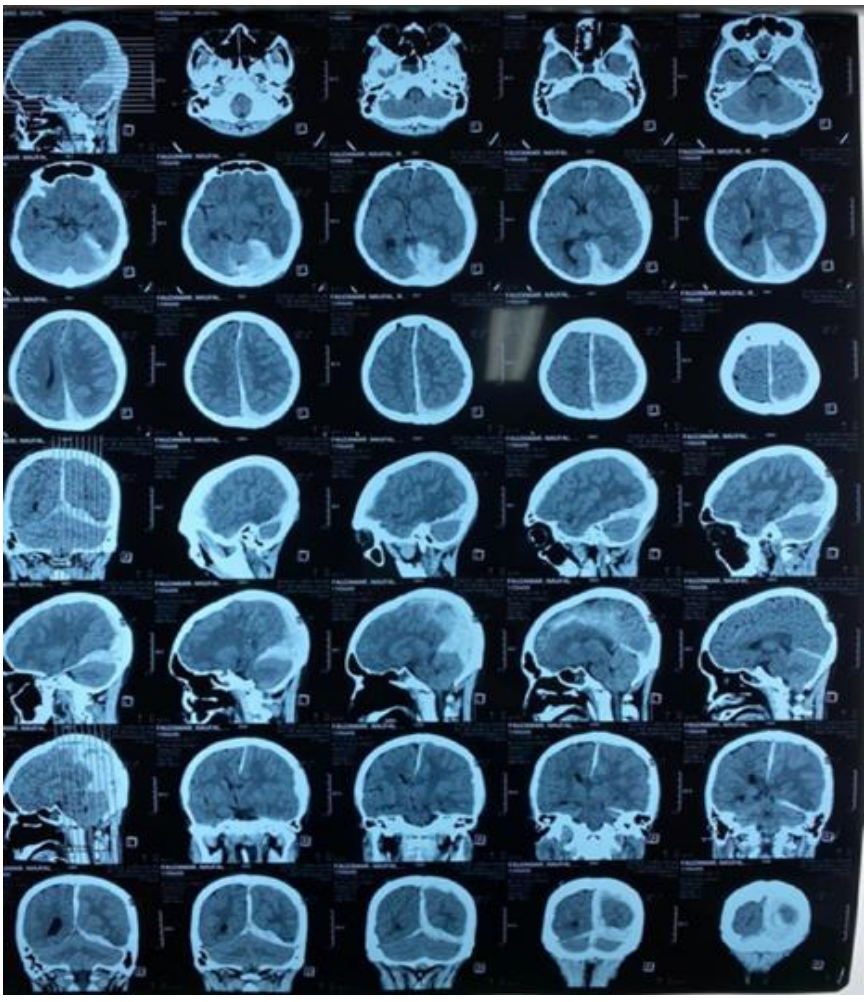

Figure 1. Preoperative CT Scan

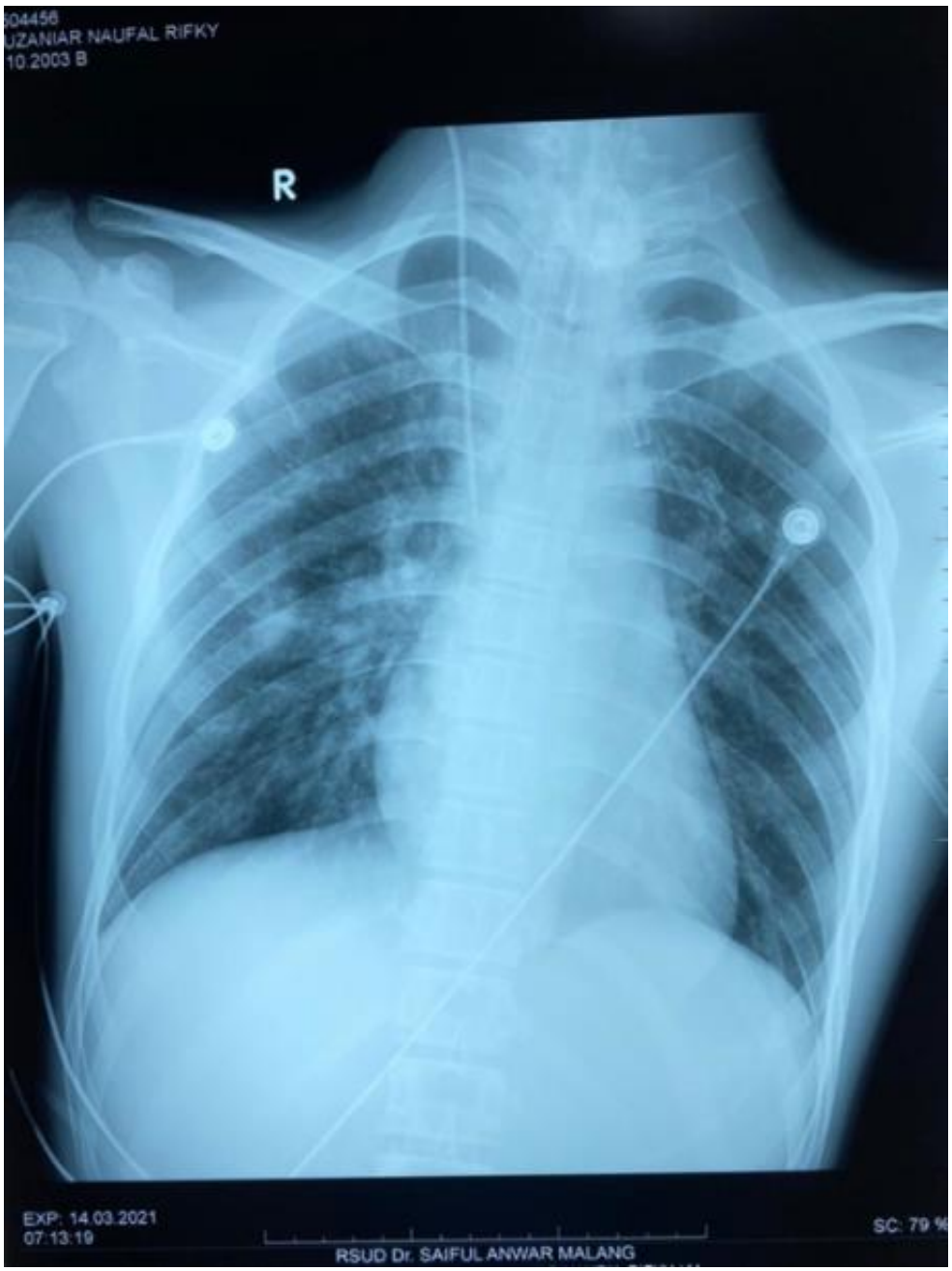

Figure 2. Preoperative chest X-ray

DISCUSSION

Intracranial bleeding is a life-threatening condition. It is bleeding within the brain parenchyma that can occur spontaneously or due to trauma. The incidence is estimated to be around 25 per 100,000 people annually. It is necessary to distinguish the types of intracranial hemorrhage between traumatic and non-traumatic. In the case

of non-traumatic bleeding, any different etiologies should be considered. In cases of intracranial hemorrhage, surgery should be conduct as soon as possible to get a better prognosis.

In this case, a 17-year-old man diagnoses with a subdural hematoma caused by trauma and hemophilia A. Ventilation control with a definitive airway and protocol for traumatic brain injury was used for initial management. Hemophilia A is an X-linked recessive disease. Women with hemophilia pass genes on to 50% of male offspring, while men with hemophilia pass genes on to female offspring[3]. Hemophilia A, the most common inherited hemostasis disorder, affects 1 in 5,000 men and accounts for 80% of cases. Hemophilia A occurs in more than 400,000 men worldwide, but mostly undiagnosed, especially in developing countries[3].

In hemophilia cases, two-thirds diagnosis confirmation is made immediately after delivery of a male newborn to a carrier mother. With the incidence of spontaneous mutations in one third of cases, diagnostic confirmation of hemophilia A continues after spontaneous bleeding or after minor trauma. Intracranial hemorrhage is the most immediate life-threatening manifestation of hemophilia A, with the potential for chronic neurological deterioration and long-term neurological sequelae[3]. Intracranial hemorrhage and soft tissues around vital areas such as the airways or internal organs remain the leading life-threatening complication. The risk of complications is 28% and leads to a third of death. Intracranial hemorrhage is the second leading cause of death and the leading cause of death associated with bleeding. In patients with severe hemophilia, 10% have intracranial hemorrhage with a mortality rate of 30%[4].

Administration of a recombinant factor VIII surrogate for the treatment of acute bleeding in patients with severe haemophilia A should be started immediately prior to completing the patient evaluation. The factor VIII replacement calculation for bleeding in severe hemophilia A is:[3]

Dose of VIII factor = target factor percentage x weight (kg) x 0,5

For severe and life-threatening bleeding, administer factor VIII to achieve 100% of the target factor VIII level. If bleeding is light to moderate, give factor VIII to achieve a target factor VIII level of 30 to 50%.

Other adjunctive pharmaceutical

therapies for hemophilia A-induced bleeding include: desmopressin, tranexamic acid, epsilon-aminocaproic acid, and factor VIII inhibitor treatment. Intravenous, subcutaneous, or intranasal desmopressin (DDAVP) is useful for treating bleeding in patients with mild to moderate hemophilia A by inducing the release of the von Willebrand factor complex and factor VIII from vascular endothelial cells. Regular prophylactic infusion of factor VIII concentrate in patients with severe hemophilia A is helpful in preventing spontaneous bleeding. Factor VIII prophylaxis aims to change severe hemophilia to a milder form by keeping the lowest factor level above 1% of normal. According to the World Federation of Hemophilia, factor VIII prophylaxis is recommended in children with hemophilia. Especially after episodes of hemarthrosis to prevent joint destruction and maintain the function of the musculoskeletal system. Patients with mild and moderate hemophilia A receive factor VIII concentrate or desmopressin[3].

On preoperative assessment, consultation with hematologist and management performed during treatment (factor replacement, blood products, etc)[5]. For major surgery to be performed on a patient with hemophilia A, the factor VIII level should be close to normal (100%) before the procedure. For patients with mild hemophilia A, it is sufficient to give DDAVP (desmopressin) infusion 30-90 minutes before surgery. DDAVP increases factor VIII levels three to five times, which can restore normal hemostasis. For patients with moderate to severe hemophilia A, correction of coagulopathy requires an infusion of factor VIII concentrates in perioperative phase. Before surgery, the presence of VIII inhibitors should also be measure, because >30% of patients with severe hemophilia A who have received factor VIII concentrates or recombinant factor VIII products develop inhibitory antibodies, rendering the concentrates ineffective[6].

Perioperative care for patients with hemophilia A and inhibitors requires of higher doses of factor VIII or agents that bypass Factor VIII in the cascade. Administration of Factor Eight Inhibitor Bypassing Activity or recombinant Factor VII (rFVIIa) is useful in the presence of severe inhibitors[7]. The half-life of factor VIII is approximately 12 hours in adults, so the infusion is repeated every 8–12 hours to maintain factor VIII levels in the blood above

50%. The peak effect of factor VIII must be considered to determine the level factor VIII to be administered and the dosing interval[6-8].

Induction of anesthesia should perform slowly, and drugs such as succinylcholine are avoided to prevent shock or contraction of the muscles, which can exacerbate hemorrhagic conditions in muscles and joints[9-10]. Peripheral infusions generally tend not to bleed excessively[18]. Central venous access, if necessary, must be confirmed by ultrasound. Intramuscular injections, difficult phlebotomy procedures, and arterial injections are avoided. Concurrent use of central and arterial catheterization results in better monitoring and patient safety[11]. In this patient central venous catheter undergo with ultrasound guiding and inserted in jugular interna.

Tracheal intubation and manipulation of the patient's airway can be life-threatening and cause submucosal bleeding. Suction in the pharyngeal area should be conduct very gently. Hypertension and tachycardia during surgery may increase the risk of bleeding. Hemodynamic conditions should be maintained close to normal. The administration of the F.VIII concentrate was continued during the surgery[5-11]. In this patient has given 4000cc preoperative and 1250cc every 6 hour post operative. Anesthetic maintenance should include reducing the risk of hypertension and tachycardia to minimize bleeding[11]. Bleeding during surgical procedures should be monitored closely. In this patient, an intubation technique with brain protection was performed to avoid a secondary brain insult caused by uncontrolled hemodynamics. After the patient was intubated, the patient was sedated with fentanyl 50 mcg/hour and midazolam 5 mg/hour.

In the postoperative phase, coagulopathy is managed with infusion of factor VIII concentrate or recombinant factor VIII products. It should be continued for up to 2 weeks to avoid bleeding that could interfere with wound healing. Postoperative monitoring is very important because hemophiliac patients are at risk for delayed bleeding after surgery (≥ 48 hours after the procedure). The patient was treated with a postoperative ventilator in the ICU due to decreased consciousness. The patient underwent weaning and extubation seven days postoperatively and further treatment was conduct in the high care

unit. The use of other postoperative analgesics such as administration of opioids, acetaminophen, and COX-2 inhibitors may be considered, whereas the use of other nonsteroidal anti-inflammatory agents is not recommended[7-13]. This patient has given factor 8 1250cc every 6 hour in intensive care unit for control the blood haemostasis.

CONSLUSION

Hemophilia A is a hereditary hemorrhagic disorder due to an inherited deficiency of factor VIII, which manifests as protracted bleeding and has complications of intracranial bleeding. The blood coagulation process is influenced by several factors, include primary homeostasis (involving platelets, vasculature, and proteins) and secondary homeostasis (thrombin, fibrinogen, fibrin). The location of intracranial hemorrhage determines the etiology of bleeding, with subdural and epidural hemorrhages due to traumatic injury while intraparenchymal and subarachnoid hemorrhages due to vascular etiology. Anesthesia management in hemophilia includes preoperative (physical examination, hematology and administration of F. VIII concentrate); Intraoperative (anesthesia, bleeding minimization, blood control and airway); Postanesthesia (administration of analgesics, concentrate VIII, transfer of the patient to the ICU for observation.

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