Hemophilic patient for emergency spinal decompression

M C Rajesh, AU Jyothis Joseph George, Neethu Asok, E K Ramdas

From Department of Anaesthesia, Baby Memorial Hospital, Kozhikode, Kerala, India

Correspondence to: Dr. M C Rajesh, Shrirang, Chevayur (Po), Kozhikode – 673 017, Kerala, India. Phone: +91-9847001136. E-mail: rithraj2@yahoo.co.in

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ABSTRACT

Hemophilia is mostly an inherited genetic disorder, caused by mutations in the clotting factor gene. With the available treatment options, life expectancy of a hemophilic patient is usually of that of the general population. Hence, it is not uncommon that they present for surgical procedures. However, hemophilic patients for the major surgical procedure are always a real challenge for the perioperative physician. We have recently encountered one such patient who was hospitalized with acute paraplegia due to a mass lesion of spine and successfully managed with the recovery of motor power. His pre-operative Factor VIII level was 0. Desmopressin nasal spray has a limited role in severe hemophilic. Our main concern was effective replacement therapy and maintenance of desired Factor VIII levels not only during surgery but also in the immediate post-operative period.

Key words: *Acute paraplegia Acute spinal decompression, Hemophilic spine*

Hemophilia is an X-linked recessive disorder with a worldwide incidence of around 1 in 5000 male births [1]. Basic defect is the absence or deficiency of clotting Factor VIII (Hemophilia A) or clotting Factor IX (Hemophilia B). Although available data are limited in India; published census show an incidence of nearly 1 in 10,000 live male births [2]. Occasionally they can present as an acquired form of the disease by the development of auto-antibodies against their own Factor VIII or in hematological malignancies [3]. Bleeding manifestations in hemophilia - A patients parallel with the decrease in Factor-VIII levels [4]. Easy bruising in the childhood, spontaneous bleeding especially in the joints and excessive bleeding following minor traumas or surgery are typical histories they narrate. Laboratory investigations usually show a prolonged activated partial thromboplastin time (aPTT), but a definitive diagnosis depends on Factor VIII assay levels.

The main challenges in handling hemophilic patients in the perioperative period are to monitor and maintain optimal Factor VIII level and detecting and dealing with cases of Factor VIII antibodies. Conventional replacement therapy will be ineffective in a case of hemophilic - A patient with high antibody titer. Alternative therapy with bypassing agentsis required in such scenarios. Routine history taking will reveal patients coagulation status and hereditary nature of the disease. Essentially managing hemophilic patients in the perioperative period involves close interaction between anesthesiologist, surgeon, hematologist, and blood bank personnel. As soon as surgery plan is fixed, the pharmacy should be notified about the expected requirement of Factor VIII.

CASE REPORT

A 60-year-old male patient of known hemophilic (Hemophilia A) presented with sudden onset of weakness of both lower limbs of 5 days duration. Started as heaviness of both lower limbs followed by difficulty in walking and retention of urine. He is hypertensive on Amlodipine Besylate 5 mg once daily. Two of his uncles had a history of hemophilia. 10 years back he underwent piles surgery, following which he had continuous bleeding. He had to be transfused with 25 units of blood, two ampoules of Factor VIII and cryoprecipitate to stop bleeding at that time. There is also the history of excessive bleeding episodes following dental extractions in childhood, which required transfusions. There was also the history of joint pains and swellings, which used to subside with bed rest and ice packs.

Clinical examination revealed Grade V power of both upper limbs and Grade 0 in both lower limbs. There was sensory deficient below 10^{th} dermatomal level bilaterally. Magnetic resonance imaging spine showed destructive marrow lesion in 9^{th} vertebral body with the possibility of neoplastic lesions such as solitary plasmacytoma or metastasis. There is a significant narrowing of the spinal canal. Pre-operative biochemical tests showed aPTT of 91 s for test and 30 s for control. Factor VIII assay showed level below 1%. Prothrombin time was within normal limits. Complete blood count and renal function tests were within normal limits. Platelet count 2.5 Lakhs/ml; Plasma fibrinogen 2.7 mg/dl. Liver functions showed elevated serum alkaline phosphatase of 116 U/L (Normal 28–74 U/L). Renal function tests, serum electrolytes within normal limits.

The patient was posted for dorsal laminectomy and decompression of mass lesion with Moss-Miami fixation. With the challenge of attaining optimal Factor VIII levels perioperative period, replacement therapy was initiated. In the absence of Factor VIII inhibitors, each unit of Factor VIII infused is known to increase levels of Factor VIII by 2 IU/dl [5]. Consequently; he was administered with Factor VIII starting 1 h before the procedure with the target of attaining Factor VIII levels at 100%. Calculated dose was given as a continuous infusion, and factor level monitored. Desmopressin nasal spray is given preoperatively at a dose of 300 μg 2 h before the procedure. Desmopressin can increase Factor VIII and von Willebrand factor levels and by their release from stored endothelial cells [6]. Von Willebrand is a glycoprotein and is necessary to carry Factor VIII in circulation [7].

Preoperatively under local anesthesia and ultrasound guidance, we have cannulated left radial for invasive hemodynamic monitoring and blood collections. Right internal jugular vein cannulated with a triple lumen 18 G jugular cannula. Routine five-lead electrocardiogram, pulse oximetry, neuromuscular monitoring, temperature probe (nasopharyngeal probe) as well as blood samplings for activated coagulation test, and arterial blood gas analysis were done during surgery. Thiopentone sodium was used for induction and atracurium used as a muscle relaxant. Fentanyl and intravenous paracetamol were used for perioperative analgesia. Synthetic antifibrinolytic tranexamic acid is given as a loading dose of 10 mg/kg and maintained at 1 mg/kg/h [8]. Surgical colleagues reported normal hemostasis during the perioperative period. The patient was reversed with neostigmine and atropine and extubated uneventfully. Factor VIII levels were monitored in the post-operative period and maintained in arranges of 80–90% with replacement therapy. The patient had an uneventful post-operative course with normal wound drain and recovery of motor function and discharged uneventfully.

DISCUSSION

The main challenge in managing the hemophilic patient for surgery is maintaining proper hemostasis. This is achieved by doing specific investigations to assess coagulation status and meticulous replacement strategies. Care should be given for proper positioning of the patient throughout the intraoperative period and avoiding pressure points. Comprehensive pre-operative assessment plan should also include inhibitor screening and assay [9]. This is all the more important when the anticipated Factor VIII levels are less than expected after replacements [10]. It is important to maintain an adequate level of Factor VIII in the extended post-operative period, till adequate tissue healing is achieved.

Re-bleeding can occur in the post-operative period after a period of hemostasis. This happens due to lysis of formed clot. Factor VIII is available in different strengths with each vial contains 250–3000 units each. Half-life of infused Factor VIII is 8–12 h. Desmopressin is useful to increase Factor VIII levels in mild and moderate hemophilics [6]. It is important for the anesthesiologist to maintain healthy communication between the lab personal, pharmacist, hematologist, and surgeon. Before committing to the perioperative care, anesthesiologist should make sure enough availability of adequate blood components and Factor VIII. For optimal support from blood banks and pharmacy, it is better to have the surgery scheduled at the beginning of the week and particularly in the morning sessions. Often overlooked but imperative aspect of the management is to monitor Factor VIII inhibitor levels, especially when the patient is on continuous replacement protocol. They are at high risk of development of antibodies [10]. There is no convincing literature to support the use of Tranexamic acid specifically in hemophilic. However, there is evidence to support its usefulness in spine surgery [11] and von Willebrand disease [12]. There is a specific recommendation of maintaining Factor VIII levels for 10–14 days with replacement therapy after performing laminectomy [13].

CONCLUSION

A hemophilic patient for spinal surgery is a real challenge; especially, when they present with acute paraplegia, warranting emergency spinal decompression. With a multidisciplinary team, approach and protocol based timely intervention in replacement therapy; these groups of patients can be safely and effectively managed. A major procedure like spine decompression requires 100% replacements of Factor VIII before the procedure. It is important to maintain Factor VIII levels in the post-operative period also till satisfactory tissue healing is achieved.

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