Original Article

Postoperative outcome in patients with hemophilia: A retrospective case series

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ABSTRACT

Background: Despite the lack of the general surgical guidelines for hemophilic patients, considering some applicable rules on preoperative, intraoperative and postoperative management of hemophilic patients who are candidate for major surgeries is sensible and even vital. Methods: This study describes a series of hemophilic patients aged 18 years and older who underwent different types of surgeries in Rasoul-e-Akram or Firoozgar hospital in Tehran between 2005 and 2015. All baseline characteristics as well as postoperative complications were collected from the hospital recorded files. Results: In total, 4 patients underwent different surgeries (one of the patients underwent surgery three times with total six surgical times). The surgical procedures consisted of gastric biopsy, synovial joint biopsy, hypophysectomy, and pectoral nerve grafting, osteotomy, and shoulder fusion. Procedural-related death was observed only in one patient. Also, preoperative blood transfusion was necessitated for only two patients. None of the major complications including local or systemic infections, deep vein thrombosis, intraoperative bleeding or readmission was reported. Conclusion: Considering proper preoperative and postoperative cares as well as timely administrating factors and blood products, the postoperative adverse events can be effectively minimized.

Keywords: Hemophilia, preoperative, retrospective

Introduction

About one percent of the general population suffers from congenital bleeding and coagulative disorders[1]. Despite low global incidence, these disorders impose a heavy financial burden on governments as well as on society through lowering patients' level of quality of life and increasing their disability[2]. Along with disease-related potential limitations, the disease management in emergent conditions such as major surgeries or needing general anesthesia has become a problematic issue [3, 4]. Hemophilia constitutes the bulk of these disorders. It has been proposed that to achieve more appropriate and safer surgical outcome in hemophilic patients, following a multidisciplinary approach with involvement of surgeons, hematologists, anesthetists, professional nurses, and even physiotherapists is potentially required [5]. In other words, despite the lack of the general surgical guidelines for such patients, considering some applicable rules on preoperative, intraoperative and postoperative management of hemophilic patients who are candidate for major surgeries is sensible and even vital. In this regard, to minimize surgical adverse outcome, preoperative preparation, intraoperative handling, and postoperative special cares should be considered [6]. The present case series describes postoperative outcome of the hemophilic patients who underwent major surgeries. The main goal of this description can help to optimize care management in such patients.

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Materials and Methods

This study describes a series of hemophilic patients aged 18 years and older who underwent different types of surgeries (abdominal, musculoskeletal, and head and neck) in Rasoul-e-Akram or Firoozgar hospital in Tehran between 2005 and 2015. All baseline characteristics including demographics, length of stay in hospital, need to transfusion, the use of coagulation factor, as well as postoperative complications (major bleeding, re-hospitalization, surgical wound or systemic infections, deep vein thrombosis, and death) were collected from the hospital recorded files and recorded at the especial checklist.

Case 1: a 49-year man diagnosed as bronchiectasis with long history of partial gastrectomy at 12 years before admission who were scheduled for gastric biopsy. The subject suffered also from hemophilia type A that was diagnosed at the age of 46 years. The patient referred to the hospital with the complaint of continuous abdominal pain at left upper quadrant site that worsened with eating and bending. In CT scan, distended loops of small bowel and proximal of colon was revealed. No postoperative complication was noted.

Case 2: A 35-year old man with hemophilia type C (lack of factor 11] who was candidate for synovial joint biopsy. The chief complaints of the patient included pain and swelling of the left ankle as well as severe weight loss (about 30 kg) since 4 months. The patient was admitted with the primary diagnosis of septic arthritis and scheduled for antibiotic therapy. Due to lack of response to treatment, synovial joint biopsy was planned leading drainage of pus and secretions. With the diagnosis of joint tuberculosis, the patient received anti-TB treatment leading hepatotoxicity and jaundice. During hospitalization, the patient received packed cell and fresh frozen plasma (FFP), however he finally died due to cardiopulmonary arrest.

Case 3: A 21-year man with mild hemophilia type A from the birth who was candidate for hypophysectomy. The patient referred with the complaint of headache and diplopia since one month that underwent hypophysectomy due to final diagnosis of growth hormone producing pit adenoma. The subject received VIII factor (1500 units twice daily for up to 5 days after surgery) plus 2 units of packed cell. No postoperative complication was reported.

Case 4: A 29-year old man with severe hemophilia type A that underwent multiple surgical procedures including osteotomy, shoulder fusion, and pectoral nerve grafting at 27, 28, and 29 years of age respectively. At the time of osteotomy and shoulder fusion, the patient received factor VIII 2500 units, half hour before operation and also 1500 units, up to three days after surgery. The patient underwent nerve grafting because of wrist drooping. During recent procedure, the patient received factor VIII, 3000 units preoperatively and also 1500 units three times a day for 48 hours. No adverse event was revealed following two surgical procedures.

In total, as shown in Table 1, procedural-related death was observed only in one patient with overall mortality rate of 16%. Also, preoperative blood transfusion was necessitated for only two patients with overall prevalence rate of 32%. None of the major complications including local or systemic infections, deep vein thrombosis, intraoperative bleeding or readmission was reported.

Results

In total, 4 patients underwent different surgeries (one of the patients underwent surgery three times with total six surgical times. With respect to the type and severity of hemophilia, one patient suffered severe hemophilia type A from birth, one of those suffered mild hemophilia type A since birth, one subject with mild hemophilia type a diagnosed at age 46 years and another one with hemophilia type C. The surgical procedures consisted of gastric biopsy, synovial joint biopsy, hypophysectomy, and pectoral nerve grafting, osteotomy, and shoulder fusion. The latter three operations were planned for one of the patients. Here, the patients are described in detail.

Discussion

As previously pointed, the main general considerations for hemophilic patients who candidate for different types of surgeries include preoperative blood packs preparation, perioperative venous access, selecting appropriate anesthetic agents, and proper postoperative cares. At the top of these considerations, employing trained healthcare providers and also related specialists is also required. Regarding preparation of appropriate venous access, it is crucial to establish reliable perioperative venous access particularly in children, young adults, and older ones[7]. Also, blood sampling should be considered preoperatively to measure specific clotting factors[8]. It has been demonstrated that the perioperative use of anti-fibrinolytic agents can enhance patients' hemostasis and thus it is recommended to inject such agents shortly before induction of anesthesia[9]. For achieving good postoperative outcome, a multidisciplinary approach is essential. In this way, employing trained nurses to provide continuing domiciliary care such as factor administration, physiotherapists to oversee the physical rehabilitation of joints and muscles, and even psychologists for psychological advices is necessary. Along with personnel-based cares, some postoperative supportive programs should be considered that among those, the use of specific clotting factor concentrates and recombinant factor products are now considered in most countries as now the treatment of choice [10-12].

In hemophilic patients, some special considerations are now programmed postoperatively according to the severity of disease. Practically, the disease is categorized as severe, moderate, or mild with the factor VIII or IX level of less than 1%, from 1% to 5%, and more than 5%, respectively. In this regard, spontaneous or traumatic bleeding may represent in severe type, while the mild to moderate forms result in excessive bleeding only in response to trauma or surgery, if not adequately treated[13, 14]. Thus, administrating factors can

effectively prevent operation-related bleeding in such patients leading appropriate postoperative outcome. However, the exact factor level required for hemostasis and the duration of clotting factor replacement after surgery remains unknown. As another problem, access to such concentrates in developing countries is practically limited.

In our case series, because of proper preoperative and postoperative cares, the surgery-related complications were not found. Also, only one death was reported because of nonsurgical reason (cardiopulmonary arrest). Also other site of bleeding must be assessed as well [15]. In other words, based on our observations, considering proper preoperative and postoperative cares as well as timely administrating factors and blood products, the postoperative adverse events can be effectively minimized. However, the lack of concentrates should be considered as an alarm for government.

Table 1: The details of hemophilic patients undergoing different types of surgeries

Case number	Age	Type of surgery	Type of hemophilia	Postoperative hospital stays (day)	Death	Infection	Intraoperative bleeding	Re-hospitalization	Deep vein thrombosis	Preoperative blood transfusion
1	49	Gastric biopsy	Type A	6	Negative	Negative	Negative	Negative	Negative	Negative
2	35	Joint biopsy	Type C	12	Positive	Negative	Negative	Negative	Negative	Positive (2U, FFP)
3	21	hypophysectomy	Type A	4	Negative	Negative	Negative	Negative	Negative	Positive (2U, PC)
4	28	osteotomy	Type A	7	Negative	Negative	Negative	Negative	Negative	Negative
4	29	shoulder fusion	Type A	6	Negative	Negative	Negative	Negative	Negative	Negative
4	30	nerve grafting	Type A	45	Negative	Negative	Negative	Negative	Negative	Negative

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