Clinics in Surgery

Case Report
Published: 13 Jan, 2021

Management of Hemostasis in Acute Aortic Dissection Type A Diagnosed with Von Willebrand Disease - Case Report

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Abstract

Background: Von Willebrand Disease (VWD), the most common hereditary bleeding disorder, arises from quantitative or qualitative defect of VWF. There are only a few report of patient undergoing cardiac surgery with VWD.

Case Presentation: A patient knows as type 1 VWD was diagnosed with Type A Aortic Dissection. He was treated prior to and after surgery with cryoprecipitate preventing coagulopathy in the absence of the von Willebrand/factor VIII concentrate.

Conclusion: Despite the absence of von Willebrand/factor VIII concentrate in our country the use of cryoprecipitate allowed our patient to undergo the aortic surgery without any complication.

Keywords: Von Willebrand disease; Aortic dissection; Bleeding

Introduction

It is known, that approximately 15 percent of the population have an abnormality of the hemostasis that results in a bleeding tendency. Of those referred for investigation, 15 to 20 percent will be diagnosed as Von Willebrand disease [1]. Von Willebrand's disease is the most common inherited bleeding disorder and has an autosomal inheritance pattern. The disease is characterized mainly by mucosa-associated bleeding and bleeding after surgery and trauma. The diagnosis is based on a personal or family history of bleeding and laboratory evidence of abnormalities in von Willebrand factor, factor VIII, or both. Affected patients have reduced levels of functional von Willebrand factor, and various types of von Willebrand's disease can be distinguished on the basis of phenotypic characteristics. The goal of treatment is to increase von Willebrand factor levels by administering desmopressin or by the infusion of exogenous von Willebrand factor–containing concentrates [2]. We describe the perioperative management of a patient with coagulopathy disease presenting for emergency operation for Type A aortic dissection.

Case Presentation

A 72-years-old man diagnosed with acute aortic dissection type A was referred in our hospital in emergency condition for emergency surgery. CT-angiography performed has revealed acute aortic dissection Type A (De-bakey type I), dissection started at the ascending aorta with a long segmental extension. There was no evidence of dissection in supra aortic vessels. Patients personal history revealed that three years ago he underwent hemorrhoidal excision, when he has had profuse hemorrhage that lasted nearly two days. Family history also revealed for coagulopathy (Figure 1). After hematological consultation we took blood tests for VWF: Ag, VWF: RCo, factor VIII activity, bleeding time, Prothrombin Time (PT) and partial thromboplastin time. In admission the patient has normal blood count. PT was normal, partial prothrombin time and bleeding time prolonged, FVIII 30%, FIX normal, VWF: Ag was 20%, VWF: RCo was 18 and VWF: RCo/ VWF: Ag was 0.9. All these findings suggested that the patient suffers from type 1 VWD. For management of hemorrhage we administered 2 h before surgery DDAVP (desmopresine) 0.3 microgram/kg intravenously diluted in 50 mL of normal saline and infused over 20 min to 30 min as test in dose. After 40 min we measure the levels of VWF and factor VIII. They were increased by two fold approximately 47% (VWF) and 66% (FVIII). Tranexamic acid 10 mg/kg iv was administrated 1 h before surgery. After that he was taken in operating room for emergency surgery. A modified Bentall's procedure was performed. Duration of the CPB was 160 min, aortic cross-clamp was 94 min, and the duration of the moderate hypothermic circulatory arrest was 29 min total postoperative bleeding was 950

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Received Date: *15 Dec 2020*
Accepted Date: *06 Jan 2021*
Published Date: *13 Jan 2021*

Citation:
Raif C, Viola C, Lako S, Sabian M, Baris U. Management of Hemostasis in Acute Aortic Dissection Type A Diagnosed with Von Willebrand Disease - Case Report. Clin Surg. 2021; 6: 3030.

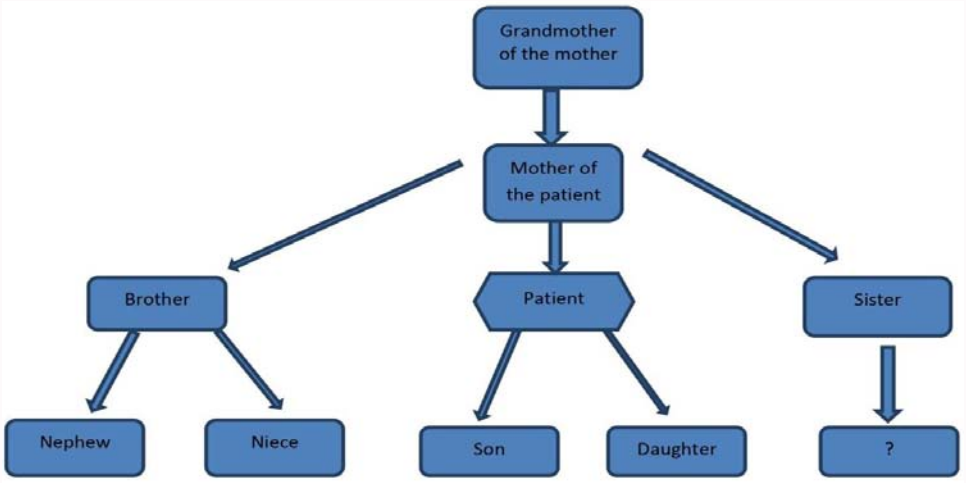

Figure 1: Family history. This members are all positive for von Willebrand disease.

ml. The thoracic drains were removed on the 6 postoperative days. On the first postoperative day we administered DDAVP 0.3 mg/kg intravenously over 30 min after 12 h and subsequent doses were switched to once daily doses for two other days to prevent tachyphylaxis and hyponatremia occurring with prolonged dosing. In the absence of von willebrand/factor VIII concentrate we transfused the patient with 8 units of cryoprecipitate (1 unite for 10 kg), 1hour before surgery and 8 units after 12 h. We continued to administer tranexamic acid 10 mg/kg every 8 h for four days. Cryoprecipitate was given on the same doses for 7 postoperative days. After surgery we administered 2 units of blood and 6 units of platelets. On the first postoperative day Factor VIII was 35%, VWF: Ag was 40%, bleeding time and a PTT prolonged. On the 7th postoperative day PT, a PTT and bleeding time were normal, FVIII 80% and VWF: Ag was 90%. Coagulation parameters were checked every day till discharge. The patient continued to recover without any bleeding complications. He was discharged from the hospital on the 12th postoperative day.

Discussion

Von Willebrand's disease is subdivided into types 1, 2, and 3. Type 1, which accounts for 70% to 80% of cases, is characterized by a quantitative deficiency of von Willebrand factor. Type 2, accounting for approximately 20% of cases, is caused by dysfunctional von Willebrand factor, resulting in a normal or reduced von Willebrand factor antigen concentration but a large reduction in von Willebrand factor function. Type 2 is further subdivided on the basis of specific phenotypic characteristics. Type 3 von Willebrand's disease is rare (accounting for <5% of cases), is the most severe form, and is caused by the absence of circulating von Willebrand factor [2]. The cornerstone of the treatment for von Willebrand's disease has been the replacement of the vW factor. But in the developing countries, where the von willebrand/factor VIII concentrate couldn't be obtainable, the sources of the vWF can be used. Cryoprecipitate is very effective in correcting the abnormalities in bleeding time associated with von Willebrand's disease. It is rich in large millimeters (fibrinogen, factor VIII and Von Willebrand factor) and is usually administered in a dose of 1 unit/10 kg. This dose will rise the factor VIII level by 15 to 20 percent, and will provide enough high molecular weight VWF to correct the bleeding time in most circumstances [1-3]. DDAVP has been used successfully for decades as perioperative non-replacement therapy for VWD. DDAVP releases factor VIII/von Willenbrand factor from storage sites. DDAVP seems most effective in type 1 and least effective in type 3 von Willebrand's disease [4]. Satisfactory response to FVIII/VWF concentrate and contraindication, FVIII/VWF concentrate was chosen as the treatment of choice to prevent peri- and postoperative bleeding. There are only a few report of patient undergoing cardiac surgery with VWD [4,5]. Its known that using FVIII/VWF concentrate at the preoperative period major procedures can be performed safely. But some circumstances when FVIII/VWF concentrate is not available like in the developing countries, these procedures can be performed safely. Despite the challenges posed by the present patient's previous hematologic history, careful preoperative evaluation and planning allowed this patient to undergo aortic surgery. In multidisciplinary approach to patients with VWD presenting for aortic surgery, close communication between anesthesits, surgeons, internists and hematologists is essential for successful management in the peri-operative period.

Conclusion

We present a case of von Willebrand type 1, diagnosed with aortic dissection undergoing surgical intervention under cryoprecipitate as the sole treatment of coagulopathy. Despite the absence of von Willebrand/factor VIII concentrate in our country the use of cryoprecipite allowed our patient to undergo the aortic surgery without any complication.

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