VIA MEDICA

CASE REPORT

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Coronary artery bypass graft in a patient with Von Willebrand disease type 1

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ABSTRACT

Von Willebrand disease (VWD), is the most common hereditary bleeding disorder (HBD) and affects approximately 1–2% of the population. Type 1 accounts for 70–80% of all cases and involves partially reduced levels of functional VWF. It is usually manifested by mild to moderate mucocutaneous bleeding. A 63-year-old patient with multivessel coronary artery disease and diagnosed with Von Willebrand's disease type 1 after a previous NSTEMI infarction (10 days earlier) was admitted to the cardiac surgery department for a planned CABG procedure. The coagulation factor values on admission to the ward were successively (VIII 15%; VWF: Ag 12%). Coronary angiography revealed MV CAD. The patient was consulted with the HEART Team. The decision was made to qualify the patient for CABG using the OPCAB technique. A detailed plan for securing the haemostasis was established. Haemate P 500/1200 and 1000/2400, a lyophilized concentrate of humans VIII and von Willebrand Factors, were secured. During surgery, LITA- LAD and Ao-RCA bridges were performed using Medtronic Starfish® Heart Positioners. In the post-operation period, no complications were noted. Plasma levels of VIII and VW factors were measured daily and antihemorrhagic prophylaxis was given accordingly to measured values. The patient was discharged on the eighth day after surgery in good condition, with the haematological recommendations. Currently, there are no guidelines for the management of patients with Von Willebrand's disease undergoing cardiac surgery. Incorrectly conducted pharmacotherapy may result in an elevated level of VW factor and additional exposure to the occurrence of acute coronary syndromes and heart attacks.

Key words: atherosclerosis, CABG, haemorrhagic diathesis, cardiac surgery, haematology

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Introduction

Patients suffering from *hereditary blood-clotting disorders* (HBD) are at particular risk of life-threatening complications if cardiac surgery is required. One of the most common HBD which affects approximately 1–2% of the population is Von Willebrand disease (VWD). It is usually manifested by mild to moderate mucocutaneous bleeding [1]. Currently, there are no guidelines for the management of patients with Von Willebrand disease undergoing cardiac surgery and the few reports on this subject encourage further research and deepening of knowledge.

Patient presentation

This study presents a case of a 63-year-old patient with unstable multivessel coronary artery disease diagnosed with Von Willebrand disease type 1 with a history of recent acute coronary syndrome (NSTEMI; 10 days earlier) who was referred to surgical coronary revascularization. LVEF was 50% and the patient received NYHA status III and CCS II. EuroSCORE II was 1.28%. Coronary angiography revealed critical left anterior descends (LAD), 80% diagonal 1 (D1) and 80% right coronary artery (RCA) narrowing (Fig 1, 2). Despite that percutaneous

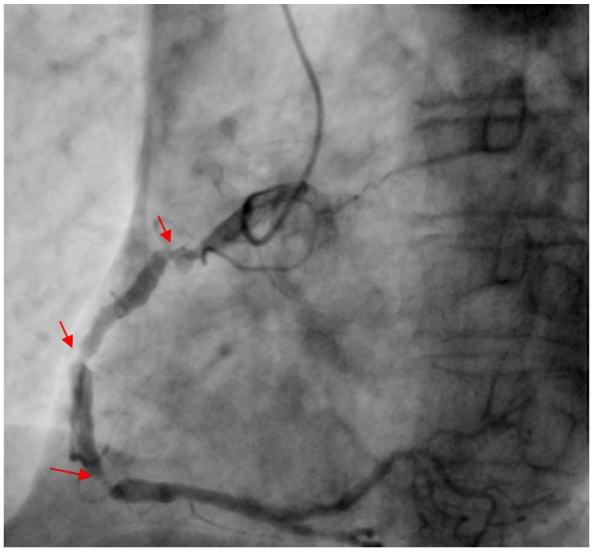

Figure 1. Coronarography of the right coronary artery with marked narrowing

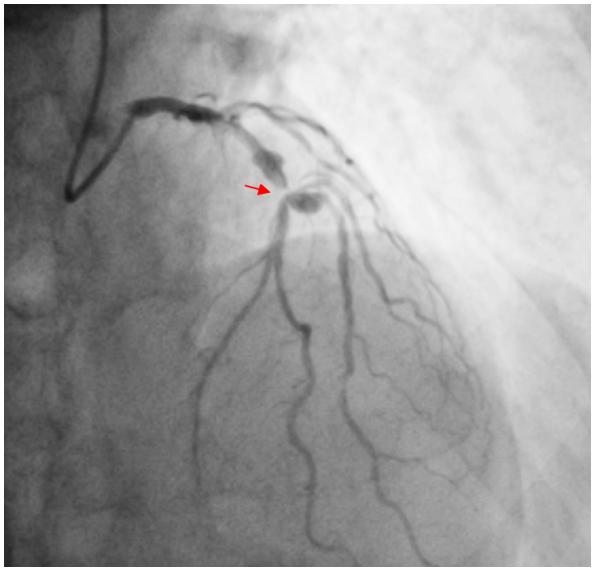

Figure 2. Coronarography of left coronary artery with marked narrowing

coronary intervention (PCI) is more secure, the patient was disqualified from PCI due to many aneurysmal lesions within the constriction and coronary arteries (D1, D2) departure. The risk of aneurysm rupture during stent dilatation was excessive. After the Heart Team consultation, the patient was referred for surgical revascularization with the preferred off-pump technique. Ten days after NSTEMI patient was admitted to the cardiac surgery department for an operation. LVEF was 50% and the patient presented symptoms of NYHA III and CCS II. The operative risk according to EuroSCORE II was calculated at 1.28%. A haematological consultation was ordered.

Preoperative anti-haemorrhagic management

The coagulation factor values on admission to the ward were successively: factor VIII — 15%; VWF: Ag — 12%. A detailed plan of securing the haemostasis, based on the lyophilized concentrate of human VIII and von Willebrand Factors (Haemate P- CSL Behring) 500/1200 and 1000/2400, was established. A preoperative treatment algorithm was implemented which included the withdrawal of acetylsalicylic acid (ASA) just before surgery and the administration of 10 doses of Haemate P 500/1200

Surgery procedure

2 hours after the start of the surgery 150 mg of Heparin was administered. ACT was 415 seconds. During the off-pump procedure two coronary artery grafts were performed using Medtronic Starfish® Heart Positioners: left internal thoracic artery to the left anterior descendent artery and vein graft to the right coronary artery. Next 2 hours later, 80 mg of protamine sulphate was injected. ACT reached 136 seconds. Prolonged, careful control of haemostasis was carried out. The surgical procedure lasted 5 hours and 40 minutes. The patient was haemodynamically stable during the perioperative period.

Postoperative anti-haemorrhagic management

In the postoperative observation, no complications were noted. Immediately, six hours after the procedure and on the first day, low molecular weight heparin (LMWH; Clexane) 40 mg was administered. Acetylsalicylic acid (75 mg) was given on the second postoperative day. Daily measurements of factor VIII and VW levels were implemented and antihemorrhagic prophylaxis was constantly adjusted to the patient's test results (Fig. 3, 4).

The patient was discharged on the eighth day after surgery in good condition. Haematologist recommended self-administration of 2 FANHDI 1000/1200 or 4 FANHDI 500/600 every third day for two weeks.

Conclusions

Taking into account numerous environmental and social factors and the increase in the incidence of cardiovascular diseases, factor into the frequent occurrence of heart attacks in patients with Von Willebrand disease. Incorrectly conducted pharmacotherapy may result in an elevated level of VW factor and additional exposure to the occurrence of acute coronary

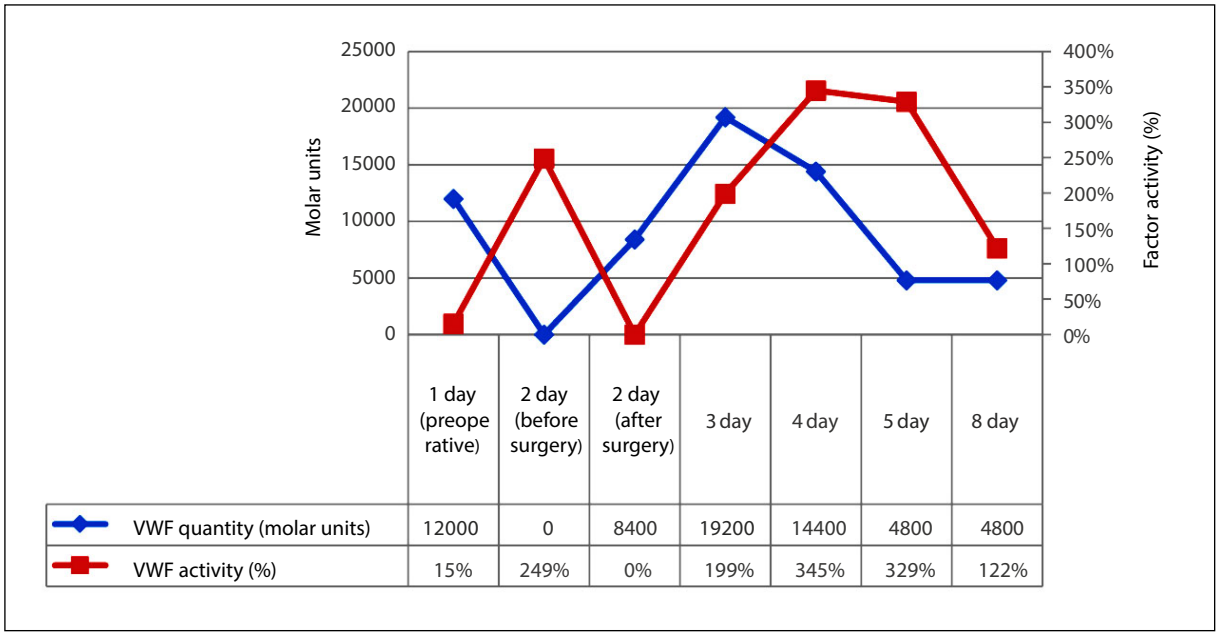

Figure 3. VW factor substitution and activity. VWF — Von Willebrand Factor

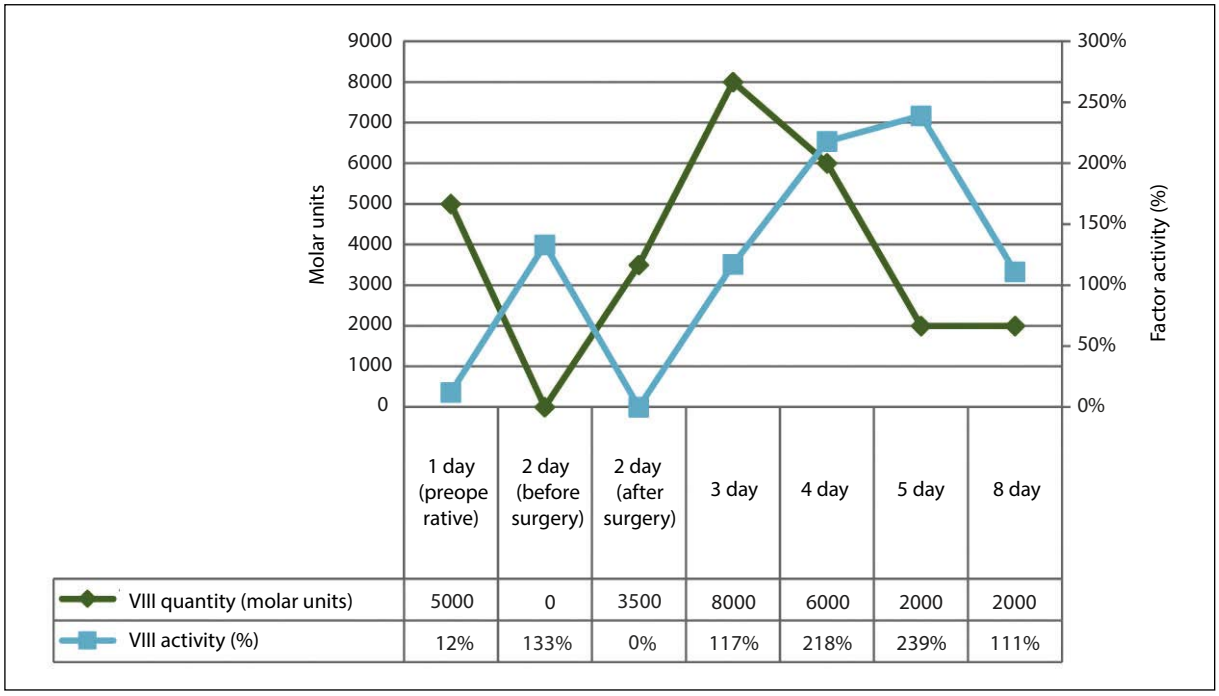

Figure 4. VIII factor substitution and activity

syndromes and heart attacks [2]. Protective measures during hospitalization do not significantly affect the risk of postoperative serious complications [3]. The main issue is the use of techniques that minimize the traumatic nature of treatments, such as off-pump coronary surgery. This reduces intra- and postoperative fluctuations in VW factor levels in relation to the values observed during extracorporeal circulation [4]. In the postoperative period, a relatively quick normalisation of the VW factor level can be observed within a week [5]. Appropriate anti-haemorrhagic management, in line with the haematologist's recommendations, additionally shortens the time needed to normalise the VW factor level and increases the safety of the operated patients.

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