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PERIOPERATIVE MANAGEMENT OF PATIENT WITH MAY-HEGGLIN ANOMALY

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ABSTRACT
The May-Hegglin anomaly (MHA) is a rare autosomal dominant hematological disorder characterized by varying degrees of thrombocytopenia, Dohle like inclusion bodies in the granulocytes and mutations of the MYH9 gene. The association between MHA and the risk of excessive bleeding during surgical procedures is unclear. We report here an experience during the administration of anesthesia to a patient with MHA who had thrombocytopenia and required Miles operation. Twenty units of platelet were transfused to correct thrombocytopenia and reduce oozing at the operation site during the intraoperative period, and intravenous desmopressin was administered in an attempt to control postoperative bleeding. After the administration of desmopressin, no further transfusions were required.

INTRODUCTION

The May-Hegglin anomaly (MHA) is a rare autosomal dominant hematological disorder characterized by varying degrees of thrombocytopenia, giant platelets and Dohle like inclusion bodies in the granulocytes [1, 2]. The pathogenesis of MHA is poorly understood, but platelet structure, function, and survival are usually normal. Thrombocytopenia occurs in 50% of patients with MHA, but severe bleeding is unusual. Some MHA patients have had a hemorrhagic tendency, which would not be wholly accounted for by the degree of thrombocytopenia [1-3]. Desmopressin has been administered to reduce the bleeding tendency in many disorders involving platelet function [4]. In addition, case reports describing the use of preventive intravenous desmopressin to reduce bleeding and transfusion of platelet concentrates in patients with MHA exist [5].

We report here an experience during the administration of anesthesia to a patient with MHA who had thrombocytopenia and required Miles operation. To reduce the operation related bleeding, Platelet concentrates (PC) and intravenous desmopressin were used.

CASE REPORT

A 77-year-old man with a known diagnosis of MHA was scheduled for surgery for rectal cancer. The patient had been diagnosed with MHA on the basis of large platelets, neutrophilic inclusion bodies that resemble Dohle bodies, positive MYH9 genetic test, thrombocytopenia-Platelet count of 50,000 / mm^3 and family history in the last year. He has had a history of easy bruising, but none of his family members had experienced a major bleeding episode or history of bleeding complications following the surgical procedure. Upon platelet function assay, the collagen/ epinephrine and collagen/ADP closure times were 207 seconds (normal: 79 - 181 second) and 107 seconds (normal: 51 - 111 second). Preoperative evaluation revealed that the patient's hemoglobin level was 12.6 g/dl, platelet count was 34,000 – 44,000/mm^3, prothrombin time (PT) INR was 0.93, activated partial thromboplstin time (aPTT) was 35.8 seconds and bleeding time (BT) was 2.0 minutes. The patient's daughter was also diagnosed with MHA in the last year. His daughter had been delivered by c-section without platelet concentrates transfusion or

complications.

Upon arrival in the operation room, the patient's blood pressure was 139/74 mmHg, heart rate (HR) was 105 beat/min and saturation spO_2 was 98%. After smooth induction with propofol and rocuronium, an arterial catheter and a venous line were placed. Blood sampling for platelet count evaluation was conducted at 30 minutes after incision because of diffuse oozing on operation sites. Laboratory data showed a platelet count of 25,000/mm^3. The patient was transfused with 10 units of PC and the platelet count was 73,000/mm^3 at 40 minutes after transfusion and 71,000/mm^3 at 2.5 hours after transfusion. Oozing at the operation site continued after PC transfusion. A total of 20 units of PC and 2 units of PRBC were transfused to the patient during the 390 minutes operation. The hemodynamics remained stable after the induction of anesthesia, with a systolic blood pressure of 90 - 139 mmHg, diastolic blood pressure of 60 – 74 mmHg and HR of 88 - 107 beat/min.

The post-operative laboratory data showed a platelet count of 138,000/mm^3 and hemoglobin level of 9.5 g/dl. The patient was transfused with 2 units of PRBC on post-operative day number two (POD #2) because hemoglobin level had decreased to 7.0 g/dl. The patient platelet count had decreased to 81,000/mm^3. On the same day, the patient received intravenous desmopressin at 0.3 µg/kg for about 30 minutes to reduce postoperative bleeding. BT conducted immediately upon completion of the desmopressin infusion was 2 minutes 30 seconds. On POD #3 and 6, the platelet count and hemoglobin were found to be 65,000/mm^3, 9.2 g/dl and 50,000/mm^3, 12.4 g/dl. After desmopressin infusion, no further transfusions were needed. The patient was able to leave the hospital after POD #15 in stable condition.

DISCUSSION

MHA is a rare genetic hematological disorder characterized by macrothrombocytopenias and mutations of the MYH9 gene, which are present in chromosomal region 22q12-13 and codes for nonmuscle myosin heavy chain IIA (NMMHC-IIA) [1,6]. May first described the anomaly in 1909 in a young asymptomatic female, and in 1945, Hegglin described the condition in a man and his son [5]. The Dohle like leukocyte inclusions in MHA are due to precipitation of myosin heavy chains in leukocytes. The exact incidence of the syndrome is unknown. MHA patients may have a range of symptoms ranging from asymptomatic to recurrent epistaxis, gingival bleeding, easy bruising to menorrhagia. MHA has not been associated with higher rates of infection [2]. MHA are frequently misdiagnosed with idiopathic thrombocytopenic purpura, and therefore have been treated with corticosteroids and splenectomy, which are ineffective. Most patients with MHA do not experience significant bleeding problems; therefore, treatment may not be required [3]. Platelet function is frequently normal despite severe thrombocytopenia. In case reports of seven deliveries in three sisters with May-Hegglin anomaly, none of the patients reported any history of abnormal clinical bleeding [7]. Moreover, none of the sisters received platelet or other blood product transfusions, even though they presented at the time of delivery with platelet counts ranging from 14,000 to 100,000/mm^3. Five epidural procedures and two spinal anesthetics were conducted among the three sisters without complication.

In the present case, even though the preoperative platelet count was only 34,000 – 44,000/mm^3, the patient was not transfused PC during the preoperative period because the results of his coagulation test (PT, aPTT and BT) were within normal limits and he did not show any bleeding problems such as epistaxis. However, PC was kept on hand in case of any bleeding problems. During surgery, we found that operative site oozing was more serious than expected and the platelet count was reduced to 25,000/mm^3. Accordingly, the patient was transfused with 10 units of PC, which resulted in the platelet count increasing to 73,000/mm^3; however, oozing from the operation site still did not stop. Moreover, the hemoglobin levels decreased to 8.5 g/dl. During the operation, the patient received a total of 20 units PC and 2 units PRBC.

There is little doubt about platelet survival in patients with MHA. Platelet survival is usually normal; however, there have been reports of shortened platelet survival [5,8]. For example, Sehbai et al. found that the platelet count increased from 56,000/mm^3 to73,000/mm^3 half an hour after transfusion with six units of platelets. They mentioned the platelet survival time in an HMA patient because the platelet count had decreased to 69,000/mm^3 four hours after transfusion [5]. The patient in the present case was not transfused with PC during the postoperative period, and his platelet count decreased from 138,000 to 111,000/mm^3 during the post operative 16 hours. These findings do not appear to be related to the platelet survival time in the MHA patient, but rather to postoperative bleeding. During the same time, the hemoglobin decreased from 9.5 g/dl to 7.7 g/dl.

Desmopressin is a synthetic analogue of arginine vasopressin and has become the drug of choice for prevention and treatment of bleeding in patients with mild hemophilia A and von Willebrand's disease [9]. Desmopressin has been shown to reduce the bleeding tendency in many disorders of platelet function [4]. Desmopressin appears to stimulate the endothelial release of factor VIII and von Willebrand factor into plasma; however, its mechanism of action in platelet disorders is still a matter of debate [5,10]. In a previous case report of the perioperative management of a MHA patient, the patient received intravenous desmopressin 0.3 µg/kg 1 hour before surgery. She then underwent temporofrontal craniotomy without any bleeding complications and the use of Platelet concentrates could be avoided [5]. In a pilot study conducted by Castaman *et al*. [11], DDAVP was found to be a safe and effective option for the treatment or prevention of bleeding in selected patient patients with

hematologic disorders. This effect was documented by the reduction of bleeding time and the cessation of bleeding by single infusion of desmorpressin. In a study conducted by Rose and Aledort [12], a nasal spray preparation of desmopressin was apparently effective at treating bleeding episodes and when used prophylactically for minor surgical procedures in several hemophilia A and von Willebrand disease.

In the present case, the hemoglobin levels decreased to 7.0 g/dl on POD #2, at which time the patient received two units of PRBC and 0.3 μg/kg of intravenous desmopressin. On POD #3, 6, and 12, the patient's platelet count and hemoglobin levels were 65,000/mm^3 and 9.2 g/dl, 50,000/mm^3 and 12.4 g/dl and 59,000/mm^3 and 12.9 g/dl, respectively. After the administration of desmopressin, no further transfusions were required and the hemoglobin levels slowly increased. Even though the clinical findings were improved after the administration of desmopressin, we cannot be sure that desmopressin was the only factor contributing to the reduction in postoperative bleeding. Laboratory data also showed that blood loss was decreased prior to the use of desmopressin. The hemoglobin levels were reduced from 9.5 g/dL to 7.7 g/dL on POD #1 and 7.7 g/dl to 7.0 g/dl on POD #2. In addition, the BT was slightly reduced to 2 minutes 30 seconds after desmopressin infusion, but BT was within the normal range during the perioperative period.

Because the patient was scheduled for a major operation and platelet function assay with collagen/epinephrine closure time was prolonged, pretreatment with PC or desmopressin during the pre-operation period may have helped reduce operation related bleeding and transfusion. The majority of patients with MHA do not experience significant bleeding problems; however, platelet preparation may be required in the case of any bleeding problems. desmopressin could be one option for the prevention and treatment of surgery related bleeding in patients with MHA.

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CONFLICT OF INTEREST:

The authors declare that they have no conflict of interest.

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