

Bernard-Soulier and Chronic Epistaxis: A Case Report

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Abstract

A 60-year-old female with Bernard-Soulier and Hemophilia A presented to the Emergency Department with epistaxis for three days. Despite an extended hospital admission with pharmacological, manual, and surgical interventions, she failed to achieve hemostasis and was discharged to a nursing facility under hospice care. This case report explores the challenges of managing a patient with chronic epistaxis complicated by multiple hematologic disorders.

Keywords: Bernard-Soulier; Epistaxis; Hemophilia A; Hematology; Isoimmunization

1. Introduction

Epistaxis is a common problem encountered in the healthcare setting. Sixty percent of the population has experienced an acute episode, while ten percent necessitated medical intervention [1]. This condition is classified based on the origin of bleeding in the nasal cavity. The anterior nasal cavity is the most common origin of bleeding due to the sensitivity of Kisselbach's plexus to drying and digital trauma [2]. Anterior nosebleeds are easily controlled with conservative measures such as nasal packing and topical vasoconstrictors (i.e., Afrin nasal spray), in contrast to posterior epistaxis, which is rare and typically requires direct intervention by artery ligation or embolization due to the potential hemorrhage that can occur [3].

Bleeding disorders can alter the course of presentation in patients with epistaxis. This relationship has been thoroughly studied in conditions such as hereditary hemorrhagic telangiectasia [4] and various familial blood dyscrasias such as Glanzmann's thrombasthenia [5]. However, the relationship between chronic epistaxis and patients with multiple bleeding disorders has not been documented.

This case explores the synergistic consequence of Bernard-Soulier and Hemophilia A on a patient with chronic epistaxis. Bernard-Soulier is a giant-platelet disorder marked by thrombocytopenia due to a defect in GP1b/IX/V, which is a platelet complex that binds to Von Willebrand factor to achieve platelet adhesion in primary hemostasis [6]. Treatment emphasizes anti-fibrinolytic therapy (such as tranexamic acid) and platelet transfusions for acute bleeding episodes. Preferably, platelet transfusions are leucocyte-depleted, and human leukocyte antigen (HLA) matched. However, if this is not feasible and platelet alloimmunization were to occur, recombinant activated Factor VII is another treatment option for a bleeding episode [7]. Hemophilia A is inherited in an X-linked recessive fashion and is defined as a deficiency of clotting factor VIII. Depending on the severity of the disease, it can lead to spontaneous intracranial or interarticular bleeding [8].

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2. Case Presentation

A 60-year-old female with a past medical history of Bernard-Soulier, Hemophilia A, thrombocytopenia, and seizure disorder presented to the Emergency Department with epistaxis and acute gingival bleeding. After the bleeding from her nose persisted for three days without resolution, the patient and her family decided that medical intervention was necessary. She denied any recent fevers, chills, hematochezia, or hematemesis.

Bloodwork in the emergency department revealed a platelet count of 14,000 mL, which is at baseline according to previous notes by the patient's hematologist. She also had moderate anemia with a hemoglobin of 8.6 g/dL. Additional labs, including a Comprehensive Metabolic Panel, activated Partial Thromboplastin Time, and Pro Thrombin/International Normalized Ratio were within normal limits. The patient was normotensive, and the remaining vitals were stable. On physical examination, the patient had generalized facial pallor and appeared fatigued. There was dried blood around the right nostril, and her mucous membranes were dry. No focal deficits were noted on neurological examination, and the patient was alert and oriented to person, place, time, and event. Cardiopulmonary and abdominal examinations were unremarkable.

Initially, she was given 500 mg of Tranexamic acid (TXA) in the emergency department. A sterile nasal packing called a Rhino Rocket was soaked in TXA solution and deployed in the right nostril. Hematology started the patient on 1g of TXA every eight hours, and if the bleeding persisted, they recommended transfusing one to two packs of regular platelets. The patient was admitted into the observation unit for further evaluation.

2.1. Hospital Course

On the third day of admission, blood continued to ooze from the right nostril. An ENT (Ears, Nose, and Throat) specialist performed cautery and inserted a new nasal packing coated with a topical antibiotic known as Bacitracin. Hematology ordered two units of pooled platelets and recommended starting recombinant activated Factor VII (Novo7) if the bleeding continued. A CT brain performed around this time showed no evidence of cerebral hemorrhage.

After being admitted for a week, blood continued to soak through the dressings in the right nostril. The patient's symptoms worsened one evening, and the nasal packings were changed thrice overnight. Hematology started the patient on 24 mcg of Desmopressin (DDVAP) every eight hours and transfused two units of normal platelets. The patient was upgraded to the progressive care unit from the observation unit. Three days later, hematology increased the frequency of TXA to three times a day and discontinued DDVAP. Additionally, one unit of packed red blood cells was transfused because the hemoglobin was down trending.

Around this time, the patient had new complaints of dull facial pain. A CT and MRI with contrast revealed allergic fungal sinusitis. According to the Infectious Disease specialists, there were no indications to start anti-fungal medications. Instead, they recommended an outpatient trial of steroids if ENT was agreeable.

The patient would go on to achieve hemostasis for approximately one week. During this time, her platelet count dropped to 2,000 mL. One unit of regular platelets was transfused, bringing the platelet levels back to 9,000 mL. TXA was titrated down to once a day, and a peripheral smear ordered by hematology showed no schistocytes or immature blast cells.

After a short resolution of symptoms, the patient experienced an acute bleed from the right nostril and the mouth. Hematology transfused one unit of regular platelets and prescribed two doses of Novo7 at 90 mcg/kg. ENT performed cautery, but the bleeding continued. The nostril was packed with an oxidized regenerated cellulose hemostat called a Fibrillar. The patient was also instructed to use Afrin and saline nasal spray twice in each nostril, three times daily. Despite the intervention by ENT, the bleeding persisted, and blood was oozing around the nasal packing. Another attempt at cauterization or the use of a hemostatic matrix called Floseal was refused by the patient. The patient's condition was refractory to the various treatments offered due to her chronic thrombocytopenia.

There was a discussion amongst the care team on whether WILATE would work better than Novo7 for the patient. WILATE is a factor replacement medication comprising Von Willebrand factor and Factor 8. Both drugs are designer medications and expensive treatment options. Ultimately, due to the lack of availability of WILATE, the consensus was to use Novo7 during episodes of life-threatening thrombocytopenia or anemia.

The patient was ultimately transferred to a tertiary care center to be evaluated by a neurological interventional radiologist. When she arrived at the tertiary care center, she was given two units of donor platelets. Before administering the second unit, she was given 10mg of IVIG to prevent isoimmunization. Her platelets after the

transfusion reached 20,000 mL, which was the highest platelet count she had in four weeks. She would receive an additional two units of donor platelets with 10g of IVIG prior to a bilateral sphenopalatine and right facial angular branch embolization procedure. Two days after the embolization procedure, the patient did not report epistaxis, her platelet count was 21,000 uL, and her hemoglobin was 11.2 g/dL. She was discharged shortly after.

3. Discussion

The patient continued to improve and followed up with her hematologist in the outpatient setting. Shortly after her appointment, the patient was re-admitted for refractory epistaxis. Similar to her initial admission, the patient's low platelets were challenging to manage due to isoimmunization caused by continuous transfusion of donor platelets [9]. She had an average platelet count of less than 10,000 mL throughout an eleven-week hospital stay. During this time, one unit of HLA-matched platelets was located and transfused, raising her platelet count to 35,000 mL and then dropping to 7,000 mL within three days. A bone marrow biopsy was inconclusive for any neoplastic process. The patient continuously refused invasive treatment options such as debridement of the nostrils or additional ligation procedures offered by interventional radiology.

After eleven weeks in the hospital, there was no long-term solution for the patient's chronic low platelets causing her refractory epistaxis. She was eventually discharged to a skilled nursing facility with hospice care.

The patient's native language is Turkish, which added an additional layer of complexity to the management of the patient. Despite using a Turkish translator, she could not recall previous diagnoses and had difficulty articulating her symptoms. She did not report any family history of bleeding disorders, making the diagnosis of her bleeding disorder difficult.

During her initial hospital admission, the patient achieved hemostasis briefly after embolization by interventional radiology. Given her medical history and underlying bleeding disorders, the embolization procedure should have been pursued earlier in the patient's admission [1]. For patients experiencing chronic epistaxis with multiple underlying bleeding conditions, such as this case, embolization as the first-line therapy for hemostasis should be investigated. The minimally invasive aspect of the procedure has a rare complication rate, and the collateral vasculature of the nose provides adequate perfusion [3]. Drugs such as Lusutrombopag, Avatrombopag, and Fostamatinib, should be explored as alternative treatment options because of thrombopoietin-receptor agonism which intrinsically increase platelet production [10]. Avatrombopag has been approved by the FDA for the treatment of chronic immune thrombocytopenia, which is characterized as the autoimmune destruction of platelets [11].

4. Conclusion

Further investigation on treatment modalities should be conducted for patients with multiple co-morbid bleeding conditions. More access to HLA-matched platelets to prevent patients from developing isoimmunization and treatments that increase the body's intrinsic platelet production such as Lusutrombopag, Avatrombopag, and Fostamatinib should be explored.

Compliance with ethical standards

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Disclosure of conflict of interest

None to declare.

Statement of informed consent

Informed consent was obtained.

Data Availability

The data supporting this study's findings are openly available on scholarly peer-reviewed journal databases.

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