

A Case of Obscure GI Bleed Associated with Non-ITP Thrombocytopenia

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Abstract

Obscure Gastrointestinal (GI) bleeding, defined by persistent or recurrent GI bleeding with source not identified on bidirectional endoscopy and small bowel radiological imaging accounts for nearly 5% of cases of GI bleeding [1]. Dieulafoy's lesion is a potentially life-threatening condition wherein persistent caliber submucosal arteriole can bleed torrentially [2]. If there is associated thrombocytopenia, it could be devastating combination. We hereby present a case of obscure overt GI bleeding (Melena) due to Dieulafoy's lesion in jejunum. She required blood transfusions for the quantum of GI bleeding. She had a rare inherited platelet disorder-Bernard Soulier syndrome [3] with non-ITP thrombocytopenia which was managed using Enteroscopy and APC of the lesion and blood products. We would like to highlight the need to follow a proper approach in managing bleeding tendency with thrombocytopenia to rule out inherited disorders and not blindly labelling it as ITP in a case of thrombocytopenia.

Keywords: Thrombocytopenia; Obscure gastrointestinal; Hemoglobin; glycoproteins

Introduction

We commonly come across cases of thrombocytopenia with or without bleeding, labelled as Idiopathic Thrombocytopenic Purpura (ITP), but we hereby describe a case with Obscure GI (jejunal) bleeding from a Dieulafoy's lesion in a patient with thrombocytopenia managed using Enteroscopy in a middle-aged female which was not due to ITP but an inherited Platelet function disorder. Proper evaluation is the key to diagnosing these rare bleeding disorders.

Case Details

A 50-year-old woman who presented with melena (black stools) and a significant drop in Hemoglobin levels necessitating a blood transfusion. A thorough investigation into her medical history revealed a background of thrombocytopenia. Importantly, there was no history of overt gastrointestinal bleeding since childhood, and the patient had only received platelet transfusions during delivery (Childbirth). Initial endoscopic examinations, including both upper endoscopy and colonoscopy, did not reveal any abnormalities.

However, upon conducting capsule endoscopy, a proximal jejunal vascular lesion with active bleeding was identified. Subsequent push enteroscopy unveiled a Dieulafoy's lesion exhibiting active ooze. At the time of assessment, the patient's platelet count was recorded at 40,000/cc. An APC (Argon Plasma Coagulation) procedure was performed on the vascular lesion as shown in Figure 1. Patient was doing well on 2 months follow up in OPD. This case could be misinterpreted as Immune Thrombocytopenia (ITP), further investigation was warranted in view of mild splenomegaly. Platelet aggregation studies played a pivotal role in arriving at an accurate diagnosis. These studies revealed a reduced response to Ristocetin. This clinical presentation raised suspicion of Bernard-Soulier Syndrome (BSS), a rare and severe bleeding disorder attributed to a deficiency in GPIb/IX/V³, a platelet complex responsible

for binding von-Willebrand factor. As demonstrated in the Figure 2, there is a key role of Glycoprotein Ib/IX in binding to VWF. It is important to note that individuals with BSS may also exhibit pseudo

thrombocytopenia due to the presence of giant platelets. Notably, the patient's responses to ADP, collagen, and arachidonic acid were within the normal range.

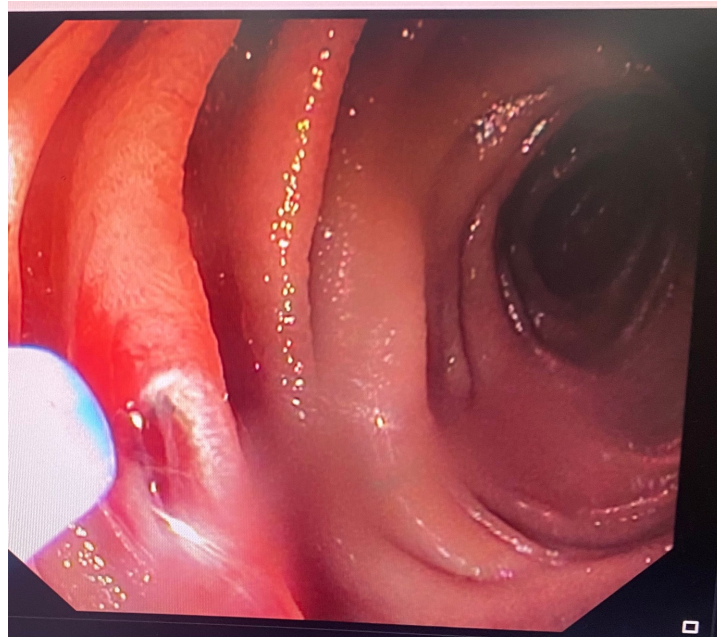


Figure 1: Bleeding spot in Jejunum-APC being done.

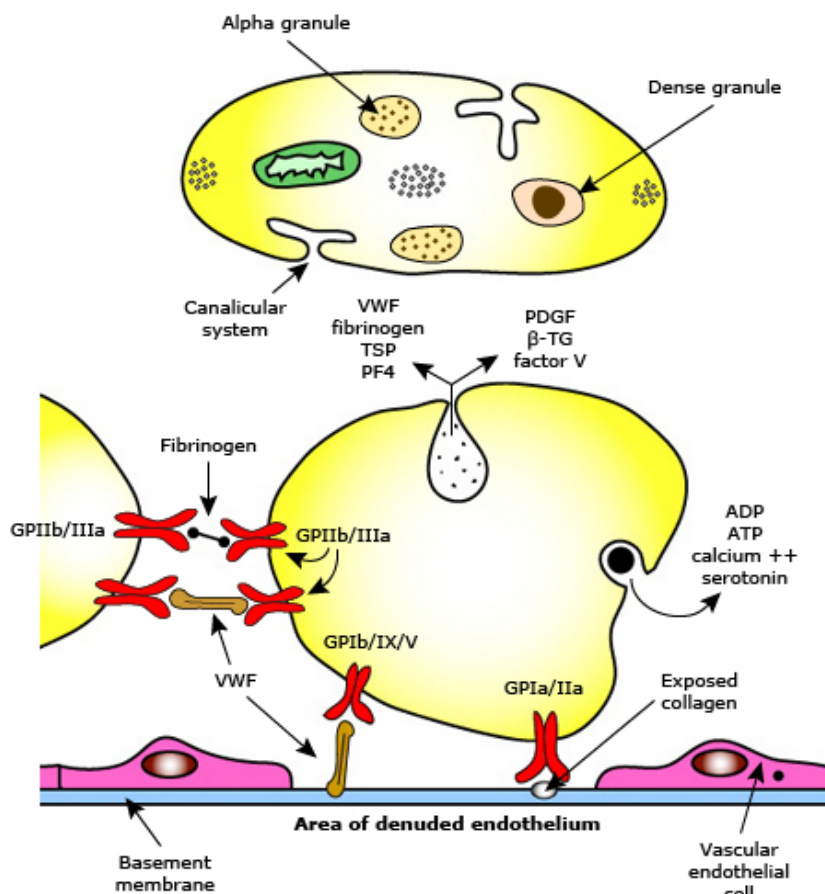


Figure 2: Platelet and the interactions. Bernard-Soulier syndrome(BSS) has defect at the level of GP Ib/IX.

Bernard-Soulier Syndrome (BSS) is an inherited platelet function disorder [3-5], Autosomal recessive in inheritance, confirmed through flow cytometry analysis of platelet glycoproteins.

It is imperative for clinicians to differentiate BSS from ITP, as the treatment approaches differ significantly. Key differentiating factors include:

- a. **Family History:** While ITP may have a familial component [3], a comprehensive family history of ITP can provide valuable insights into the diagnosis.
- b. **Response to First-Line Treatment:** Patients with BSS typically do not respond to first-line interventions such as Intravenous Immunoglobulin (IVIG) and steroids [3-5], in contrast to ITP patients who may show improvement with these treatments.
- c. **Diagnostic Tests:** Both light aggregation studies and flow cytometry [3-5] are essential diagnostic tools in distinguishing between the two conditions.

Conclusion

In conclusion, this case highlights the importance of management of obscure GI bleed with a stepwise approach and

also underscores the importance of an appropriate diagnostic approach when encountering patients with bleeding disorders and thrombocytopenia. The differentiation between Bernard-Soulier syndrome and immune thrombocytopenia is pivotal for selecting the most appropriate treatment strategy. The line of treatment varies among the two different disorders.

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