REVISTA ESPAÑOLA DE
ENFERMEDADES DIGESTIVAS
The Spanish Journal of Gastroenterology

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DOI: 10.17235/reed.2022.9081/2022
Link: PubMed (Epub ahead of print)

Please cite this article as:
Jiménez Castillo Raúl Alberto , Benavides-Salgado Daniel Eduardo, Jáquez-Quintana Joel Omar, González González José Alberto, Cortez Hernández Carlos Alejandro, Náñez-Terreros Homero, Maldonado-Garza Héctor Jesús. Glanzmann thrombasthenia: an uncommon cause of acute upper gastrointestinal bleeding. Rev Esp Enferm Dig 2022. doi: 10.17235/reed.2022.9081/2022.

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Glanzmann thrombasthenia: an uncommon cause of acute upper gastrointestinal bleeding

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Conflict of interest: none
Funding source: none
Acknowledgment: None
Type of manuscript: Letter
Word Count: 394 words.

Keywords: Gastrointestinal bleeding. Glanzmann thrombasthenia.

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Dear Editor,

The major function of platelets is to contribute to hemostasis. If an impairment in their production and/or function occurs, abnormal bleeding can develop.

An 18-year-old male presented to our hospital after four episodes of hematemesis. His medical history was relevant for Glanzmann thrombasthenia diagnosed during early childhood. On initial examination, he appeared pale and with normal blood pressure.

His complete blood count included a hemoglobin concentration of 11.0 g/dL, additional laboratory tests were within the normal ranges. The initial approach consisted of a high dose of proton pump inhibitors. Hours later, esophagogastroduodenoscopy revealed diffuse oozing bleeding from gastric mucosa with no other visible lesions such as peptic ulcers or varices, (Figure 1). We promptly began anti-fibrinolytic therapy and transfusion of platelet concentrates. No other episode of gastrointestinal bleeding was documented and we decided patient´s discharge.

Glanzmann thrombasthenia is a rare platelet function disorder with an incidence of one per million people (1). The mainstay in its pathogenesis is a genetic defect in ITGA2B (GTIIb) or ITGB3 (GTIIIa) genes on chromosome 17 (2), leading to a deficiency in platelet membrane glycoprotein (GP)IIb/IIIa, resulting in impaired platelet aggregation. Glanzmann thrombasthenia is characterized by a normal platelet count and morphology, platelets tend to be refractory to aggregation but do agglutinate in the presence of ristocetin.

Clinical manifestations range from epistaxis, gingival bleeding, and menorrhagia to hematuria, central nervous system hemorrhages, and severe gastrointestinal bleeding, which are less frequently observed (3).

An initial evaluation in patients with gastrointestinal bleeding and this platelet disorder did not differ from the general population and treatment consists of prompt achievement of hemodynamic stability. In order to know the etiology, further evaluation with esophagogastroduodenoscopy is mandatory. Source of bleeding is identified in the

majority of cases, but interestingly, in our patient, a specific lesion was not seen.

Treatment strategies in Glanzmann thrombasthenia focus on a prophylactic approach prior to high-risk procedures and supportive treatment in response to hemorrhages. Tranexamic acid is the most frequent therapy for mild to moderate hemorrhagic events and platelet transfusion is reserved for severe bleeding episodes. Recombinant human activated factor VII was approved in 2004 and is mainly reserved for those who cannot be treated with platelet transfusions (4).

Prognosis in Glanzmann thrombasthenia is derived from case series studies and despite its singularity, outcomes tend to be good with current treatment strategies. Our patient was successfully treated according to current recommendations.

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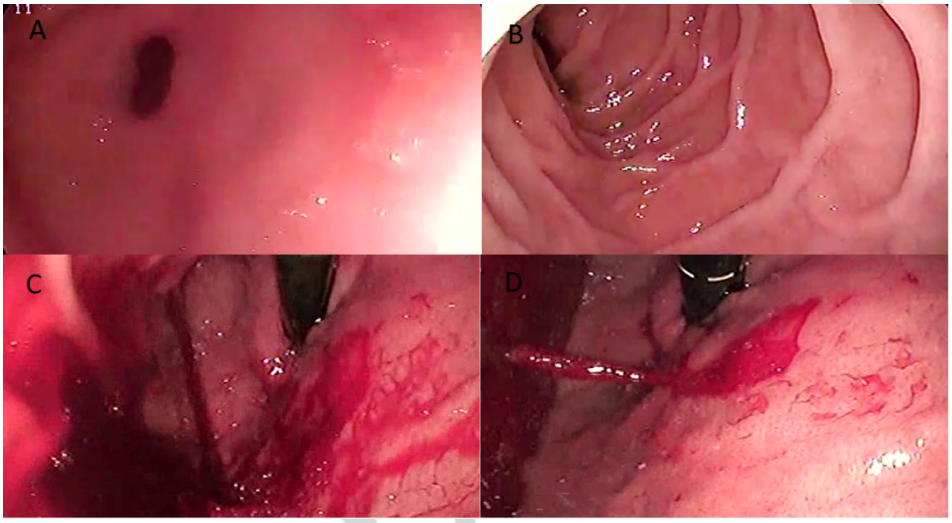

Figure Legends

Figure 1 A) Normal gastric antrum B) The duodenal bulb and the second portion of the duodenum without alterations. C) Esophagogastroduodenoscopy revealed an oozing hemorrhage in the gastric fundus. D) After a clean-up of the gastric fundus, active oozing hemorrhage recurred.