Case Report

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A RhD-Negative Girl with Dieulafoy's Lesion in the Jejunum Presenting with Life-Threatening Gastrointestinal Bleeding: A Case Report

Kai-Jun Hwang[1,2], Yu-Tsun Su[3,4], Po-Jui Ko[5], Kai-Jen Lin[6], Ching-Chung Tsai[3,4,]*

A 13-year-old girl fainted at school due to sudden massive gastrointestinal (GI) bleeding and presented with bloody stools. She had no history of systemic disease, bleeding disorders, or recent abdominal pain. An immediate panendoscopy revealed negative results. Due to persistent GI bleeding after admission, emergent RhD-negative blood was required to be transfused because of impending hypovolemic shock. However, RhD-positive blood was used because of the lack of RhD-negative blood. Urgent computed tomography angiography revealed contrast medium leakage in the small intestine of the left upper abdomen. Therefore, an intraoperative enteroscope was used to find a protruding lesion with fresh blood spurting, nearly 150 cm below the ligament of Treitz inside the jejunal lumen. Intestinal segmental resection was performed, and the patient was discharged a week later under stable conditions. Dieulafoy's lesions were confirmed by pathology. We report this case to remind physicians that Dieulafoy's lesion in the jejunum can cause massive painless and lethal bleeding in the lower GI tract.

Key words: Dieulafoy's lesion, gastrointestinal bleeding, jejunum

Introduction

Generally, hematochezia is caused by lower gastrointestinal (GI) bleeding. The most common causes of lower GI bleeding include anal fissure, hemorrhoids, colonic polyps, infectious enterocolitis, inflammatory bowel disease, intussusception, Meckel's diverticulum, and angiodysplasia.[1] Massive painless lower GI bleeding is usually caused by Meckel's diverticulum and angiodysplasia.

Dieulafoy's lesion is a rare GI vascular abnormality characterized by a large-lumen arteriole, originating from the submucosa layer, eroding into the mucosal layer of the GI tract.[2] Predisposing ulcers, surrounding aneurysms, or intrinsic mural abnormalities usually cannot be

From the [1]Department of Pediatrics and [2]International Aesthetic Medical Center, E-Da Dachang Hospital, I-Shou University; [3]School of Medicine for International Students, College of Medicine, I-Shou University; [4]Department of Pediatrics, [5]Department of Pediatric Surgery and [6]Department of Pathology, E-Da Hospital, I-Shou University, Kaohsiung, Taiwan.

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* Address reprint request and correspondence to: Ching-Chung Tsai, Department of Pediatrics, E-Da Hospital, No. 1, Yida Road, Jiaosu Village, Yanchao District, Kaohsiung City 824005, Taiwan
Tel: +886-7-615-0011 ext. 251295, E-mail: u101130@gmail.com

detected. Dieulafoy's lesion occurs more often in men over the age of 50 and is extremely rare in children.[2-4] Herein, we report a thirteen-year-old girl presenting with massive painless GI bleeding complicated by hypovolemic shock due to Dieulafoy's lesion in the jejunum.

Case Report

This 13-year-and-11-month-old girl was relatively healthy prior to current hospitalization. Massive hematochezia accompanied by syncope occurred while the patient was in school. She was quickly transferred and admitted to our pediatric emergency department (PED). Her consciousness was clear, with pale conjunctiva and pale lips. She had no history of systemic disease, bleeding disorders, or recent abdominal pain. Her vital signs were relatively stable when admitted to PED (body temperature, 36.5˚C; blood pressure, 95/63 mmHg; heart rate, 80 beats per minute; respiratory rate, 20 cycles per minute). No hepatomegaly, splenomegaly, petechia, or ecchymosis was observed in her body. In addition, her previous menstrual cycle ended approximately 3 weeks ago. Her initial laboratory examinations revealed the following: white blood cell count, 95,000/μL; hemoglobin (Hb) level, 11.1 g/dL; hematocrit, 31.4%; platelet count, 180,000/μL; neutrophil, 74.8%; eosinophil, 0.6%; lymphocyte, 19.0%; basophil, 0.2%; monocyte, 5.4%; prothrombin time, 12.8s; activated partial thromboplastin time, 30.8s; and stool routine, occult blood - 4+, mucus 1+, and pus, 1+. Owing to hematochezia and hypotension, fluid resuscitation with normal saline and hydroxyethyl starch was administered. Panendoscopy revealed no obvious peptic ulcers or active bleeders. Tachycardia and hypotension were observed owing to persistent hematochezia after admission. Hence, fluid challenge and blood transfusion with type A Rh-negative packed red blood cells (pRBC) were prescribed immediately. Unfortunately, due to continued hematochezia and low Hb (6.8 g/dL), type A Rh-positive whole blood was transfused, and Rh immunoglobulin was prescribed owing to Rh-negative blood shortage at the Kaohsiung Blood Bank. Moreover, emergent computed tomography angiography revealed local extravasation of contrast medium within the small bowel loop over the left upper abdomen (Fig. 1). Exploratory laparotomy and intraoperative enteroscopy were performed swiftly to identify the active bleeding. One 0.5 cm × 0.5 cm papillary protruding lesion with blood consistently squirting was noted 150 cm below the ligament of Treitz (Fig. 2), which was probably in the jejunal lumen. Segmental resection of the jejunum and end-to-end anastomosis were performed to stop the bleeder. Later, a pathological report showed ectatic arterioles in the submucosal layer, with thickened vessel walls (Fig. 3). Thus, the above findings are consistent with Dieulafoy's lesions. The patient's hemodynamics were secured after the emergent operation. The patient was then discharged a week later.

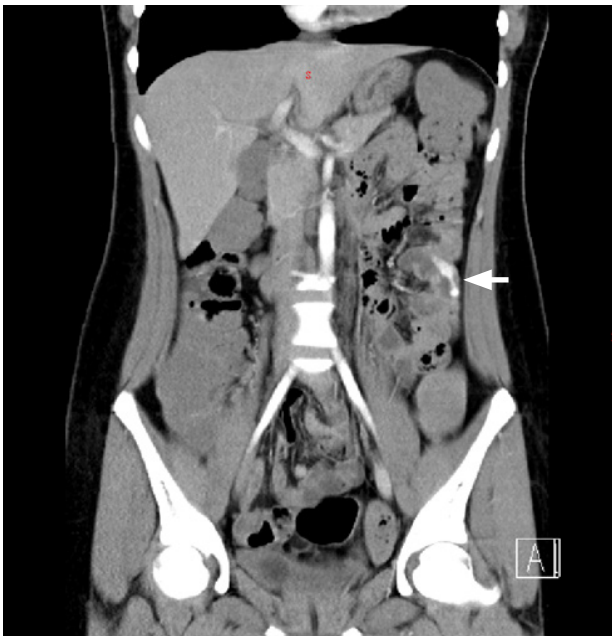

Fig. 1 Coronal view of computed tomography angiography revealed local extravasation of contrast media within the small intestinal loop (arrow) over the left upper abdomen.

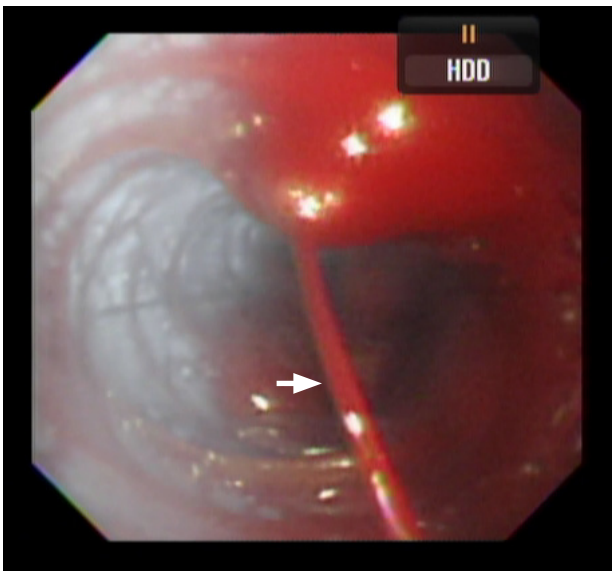

Fig. 2 Intraoperative enteroscope showed there was one 0.5 cm × 0.5 cm papillary lesion protruding into the jejunal lumen, with blood spurting (arrow) near 150 cm below the Treitz ligament.

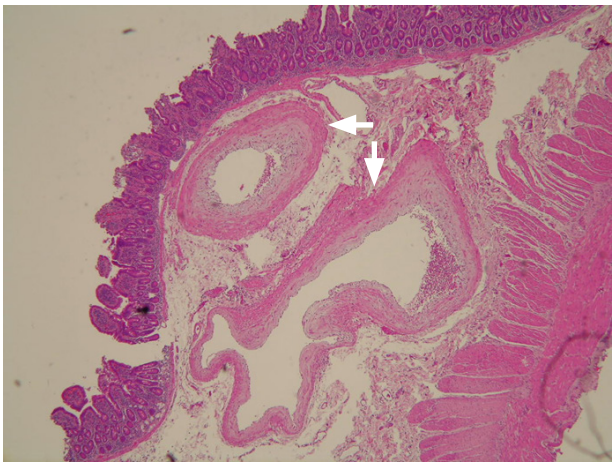

Fig. 3 Pathology (100× and Hematoxylin and Eosin staining) revealed ectatic arterioles with thickened vessel walls (arrow) were present in the submucosa layer.

Discussion

The exact etiology of Dieulafoy's lesions remains unknown. Reports have indicated that Dieulafoy's lesion may be related to the use of nonsteroidal anti-inflammatory drugs and antiplatelet drugs. It is associated with chronic diseases, such as hypertension, chronic kidney disease, and diabetes mellitus.[3] Only approximately 1% – 2% of acute GI bleeding is caused by Dieulafoy's lesion. The most common site of Dieulafoy's lesion in the GI tract is the lesser curvature of the stomach, particularly near the gastroesophageal junction. Other than stomach lesions (approximately 71%), Dieulafoy lesions can also be found in the duodenum (approximately 15%), esophagus (about 8%), and other parts of the GI tract lumen (about 6%).[5] In children, a study has reported that the stomach is the most common site of Dieulafoy's lesion, accounting for approximately 57.1%, followed by the small intestine (17.8%), colon (14.3%), duodenum (10.7%), and ileum (7.1%).[6] The clinical manifestations of Dieulafoy's lesion are hematemesis, melena, or intermittent massive gastrointestinal bleeding. Early diagnosis, fluid resuscitation, and blood transfusion are required to prevent traumatic hypovolemic shock. It can be a challenge for physicians to make an efficient diagnosis owing to variable lesion sites.[5]

Panendoscopy, enteroscopy, and coloscopy can be used as diagnostic tools and therapeutic treatments for Dieulafoy's lesions. Usually, an active bleeder can be located using an endoscope because the blood squirting is obvious. Unlike peptic ulcers, there are no surrounding ulcers in Dieulafoy's lesions.[7] Some endoscopic procedures can also be used to terminate bleeding, such as focal adrenaline injection, hemostatic clips, and band ligation.[5] If these procedures are ineffective, vascular embolization can be considered.[2] Dieulafoy's lesions are uncommon in the small intestine. Although enteroscopy can be used to diagnose lesions in the small intestine, it has some limitations. As in our case, intraoperative enteroscopy was an alternative method for making a definitive diagnosis. Delayed management of Dieulafoy's lesions may lead to a high mortality rate.[5] Thus, intraoperative enteroscopy is a method for early diagnosis to identify the active bleeder in the GI tract and remove the lesion surgically.

RhD-negative pRBCs and low-titer group O whole blood are scarce resources at most

hospitals. A previous study revealed that only 10% of pRBCs are RhD-negative.[8] Immediate blood transfusions can reduce mortality in patients with massive gastrointestinal bleeding. If RhD-positive blood is transfused into RhD-negative recipients, it will cause RhD-alloimmunization in the recipient's body. Studies have revealed that RhD-negative patients receiving RhD-positive blood transfusion in the event of accidental trauma or emergent surgery have approximately 11% – 50% RhD-alloimmunization rate.[9] RhD-alloimmunization in women of childbearing potential (FCP) may cause hemolytic disease of the fetus and newborn (HDFN) during pregnancy. Modern advanced obstetric medical technology has made HDFN a correctable disease. A study has shown that RhD-negative FCP who received RhD-positive blood transfusions in an accidental trauma patient had developed alloimmunization, and the overall rate of intrauterine fetal demise was approximately 0.3% if RhD-positive fetuses were carried.[10]

In conclusion, since Dieulafoy's lesion is an extremely rare and critical condition in children, pediatricians should consider this diagnosis when encountering massive painless GI bleeding. If a panendoscope or colonoscope yields negative results, a high index of suspicion is required for diagnosing Dieulafoy's lesion in the small intestine, and intraoperative enteroscopy could be an efficient diagnostic tool in this lethal event for early diagnosis and prompt management.

Author Contributions

KJH, CCT and YTS have contributed to collecting the clinical information and data. PJK, CCT and KJL have contributed to providing the relevant images. KJH and YTS collected references. KJH has contributed to drafting the manuscript. CCT has contributed to revising the manuscript. All authors have approved the final version of this manuscript.

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Institutional Review Board Statement

Case report is exempt from formal ethical approval at Institutional Review Board in E-Da Hospital.

Informed Consent Statement

Informed consent was obtained from all subjects involved in the study.

Data Availability Statement

Not applicable.

Conflicts of Interest

The authors declare no conflict of interest.

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