

Life-threatening bleeding for a large cameron ulcer. A novel description of a tailored-surgical strategy: report of a case and literature overview

D. BISOGNI¹, A. VALERI¹, L. TALAMUCCI², R. MANETTI², A.B.F. GIORDANO¹,
M. ARDU¹, R. NASPETTI², P. PROSPERI¹

SUMMARY: Life-threatening bleeding for a large cameron ulcer. A novel description of a tailored-surgical strategy: report of a case and literature overview.

D. BISOGNI, A. VALERI, L. TALAMUCCI, R. MANETTI, A.B.F. GIORDANO, M. ARDU, R. NASPETTI, P. PROSPERI

Hiatal hernias (HHs) are usually divided into two main groups: sliding and para-esophageal (torsional) ones. Sometimes patients presenting HHs experience progressive anemia, whereas rarely an acute anemia with melena or hematemesis can occur. In such cases a Cameron ulcer should be suspected and a careful esophago-gastro-duodenoscopy (EGDS) with a meticulous inspection of the mucosal folds along the neck of the hernia is the best examination in order to find out the ulcer itself. In front of massive hemorrhage due to a Cameron erosion, the first aim should be the control of the bleeding itself, in order to re-

establish hemodynamic stability. The Authors report the case of a 72-year-old man presenting a severe bleeding secondary to a large Cameron ulcer in a para-esophageal hiatal hernia. Firstly, a combined medical-endoscopic therapy was tried; the patient underwent transfusions of pooled red blood cells and endovenous anti-acid therapy combined with an operative endoscopic treatment; unfortunately this initial approach failed, therefore the patient was referred to surgery. The surgeons realized a minimally invasive atypical gastric resection associated with the HH repair; the post-operative course was uneventful and no other rebleeding episodes occurred. The urgency treatment of a life-threatening bleeding for Cameron ulcers remains a very challenging problem as no univocal and standardized recommendation has been described in literature since now. In this case-report the Authors make an overview of the current literature on the treatment of Cameron ulcers, describing a novel surgical technique for massive upper gastro-intestinal bleeding secondary to these lesions.

KEY WORDS: Large Cameron ulcer - Life-threatening bleeding - Endoscopic treatment - Minimally invasive atypical gastric resection.

Introduction

Hiatal hernias are usually divided into two main groups: sliding and para-esophageal (torsional) ones (1, 2). Considering the increasing rate of EGDS performed, HH has become one of the most common endoscopic finding (1). Even though usually asymptomatic, thereby constituting incidental findings, sometimes complications occur: gastroesophageal reflux disease, dysphagia, acute or chronic bleeding, and ulcer formation (3, 4). Complications of tor-

sional hiatal hernia are quite rare, but often severe (4-6); the most serious one is a massive bleeding, which is usually due to a Cameron ulcer, a benign linear gastric ulcer located at the site of the diaphragmatic constriction in patients with HH (4-6). Because of the rarity of these lesions, the interpretation of available information related to the treatment of severe bleeding secondary to Cameron erosions is confounding and, unfortunately, univocal indications are lacking (6-8). The Authors present the unusual case of an atypical gastric resection for a refractory hemorrhage due to a large Cameron ulcer (type Forrest 1A) in a patient with a para-esophageal HH, initially treated with an operative endoscopic approach. During the hospital course other bleeding episodes occurred, leading to severe anemia requiring pooled red blood cells transfusions. Subsequently the patient was referred to surgery. A minimally

¹ Emergency Surgery, Emergency Department, Careggi Teaching Hospital, Florence, Italy

² Interventional Endoscopy, Department of Oncology and Robotic Surgery, Careggi Teaching Hospital, Florence, Italy

Corresponding author: Damiano Bisogni, e-mail: bisognifelice@libero.it

© Copyright 2019, CIC Edizioni Internazionali, Roma

invasive atypical gastric resection with a mechanical stapler was accomplished, together with HH repair. The patient was discharged on the ninth post-operative day and no further bleeding episodes occurred. After a comprehensive review of the literature, this report appears as one of the first describing a gastric resection for massive upper gastro-intestinal bleeding secondary to a large Cameron ulcer.

Case report

A 72-y.o. caucasian male was admitted to first AID for melena and hematemesis. The patient complained weakness, dyspnea, palpitations, diffuse bone pain, fatigue and light-headedness for the past five days. His past medical history was significant only for arterial hypertension, HH, overweight and benign prostatic hyperplasia. His past surgical history was insignificant, since he had not undergo any surgical operation. Social history was significant for occasional tobacco smoking, without any alcohol use. His current medications included anti-hypertensive drug and Tamsulosin. He denied any allergy. A cursory physical examination in the Emergency Department was consistent with his symptoms, since he was tachycardic, pale, sweaty and presented severe conjunctival pallor. A more accurate examination immediately revealed severe hemodynamic instability, with the following values: arterial pressure 90/55 mmHg, cardiac frequency 93 bpm, rate blood oxygen 80% and hemoglobin concentration 7,2 g/dL. Liver and kidney function tests appeared within normal ranges.

The patient was immediately transfused with 4 units of packed red blood corpuscles. Since a gastrointestinal source of bleeding was suspected to be the more likely hypothesis, a contrast enhancement CT-scan and a subsequent EGDS were performed, revealing a large bleeding ulceration of about 5 cm size in diameter (Type Forrest 1A) located along a gastric mucosal fold at the diaphragmatic impression associated to a torsional HH (Figure 1): the whole situation was consistent with a large bleeding Cameron erosion. During the EGDS the active source of hemorrhage was temporarily stopped through the positioning of three hemostatic clips (Boston® clips) and local injection of cyanoacrylate; at the end of this interventional endoscopic proce-

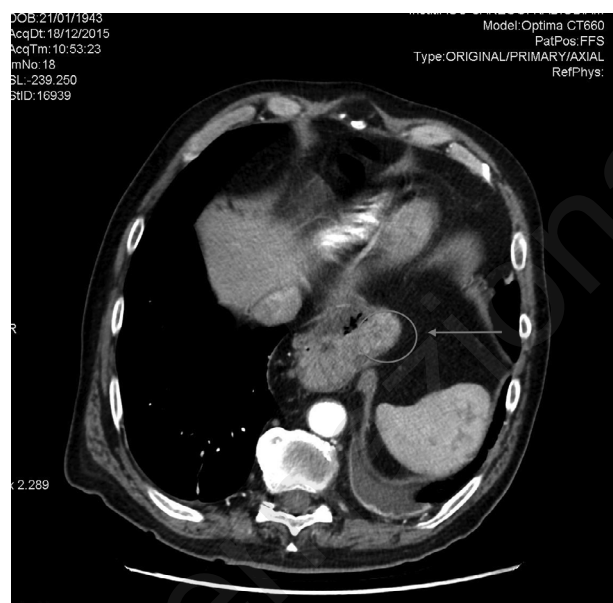


Figure 1 - CT-scan image showing the Cameron ulcer at the diaphragmatic impression with a para-esophageal hiatal hernia. In the image, the bleeding ulcer appears hyperdense compared to the gastric wall.

dures a biopsy was performed. Within the first 36 hours other severe episodes of melena and hematemesis occurred; therefore, the patient was evaluated by a multidisciplinary team and a surgical approach was considered the best choice to stop definitely the bleed. The operation was performed by two over twenty years experience surgeons. In the operatory theatre, a pneumo-peritoneum was performed according to open laparoscopic access with the Hasson's trocar and four other trocars were introduced. First of all, some adhesions between diaphragm and stomach were cut, thus exposing the Laimer-Bertelli lamina and allowing to retrieve the stomach from the thorax into the abdominal cavity. Subsequently an endoscopically-guided atypical gastric resection with a mechanical stapler was performed, followed by a gastric wall reinforcement through interrupted sutures. A careful hemostasis control was done, and a 19 French Blake tube drainage was placed into the Winslow foramen. The macroscopic aspect of the removed gastric portion seemed to be benign, thus reinforcing the Cameron ulcer hypothesis (Figure 2). Postoperatively, the patient was admitted to the intensive care unit (ICU). On second post-operative day the patient was discharged from ICU and admitted in the general ward. On the fifth post-operative day, a barium-



Figure 2 - Internal aspect of the gastric resected area, including the large Cameron erosion, as it was opened after surgery.

contrast X-rays was performed, showing a normal gastric wall impregnation without any leakage (Figure 3), therefore the patient tolerated a soft diet on the sixth post-operative day. He was discharged on the ninth post-operative day without any other re-bleeding episode occurring; the histological exam of the ulcer confirmed the benign nature of the lesion

(nonspecific inflammation of mucosa and submucosa associated with large epithelioid cells), so that, considering the clinical history of the patient and the histological findings, the diagnosis of Cameron ulcer was finally confirmed (Figures 4, 5).

In the following weeks a control EGDS revealed a normal aspect of the stomach along the surgical suture.

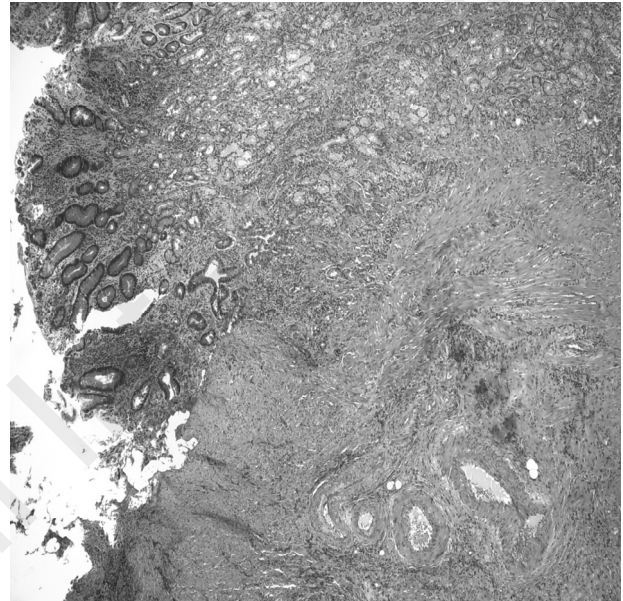


Figure 4 - Histological image of the gastric ulcer (hematoxylin-eosin x20).

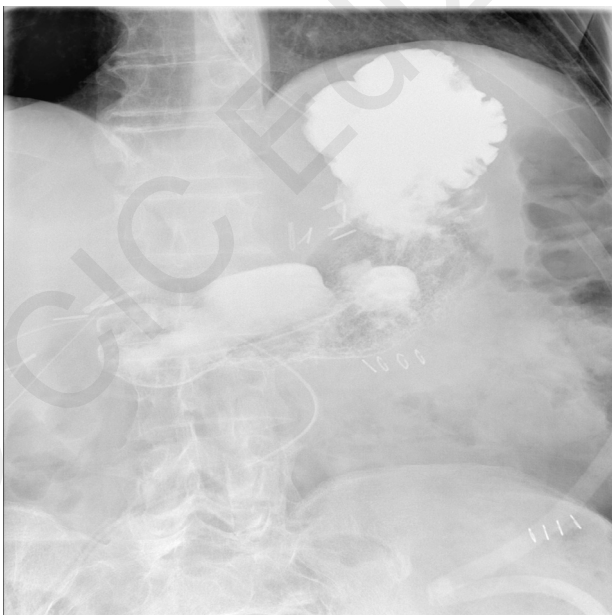


Figure 3 - Barium-contrast X-rays control showing a normal gastric wall impregnation without any leakage.

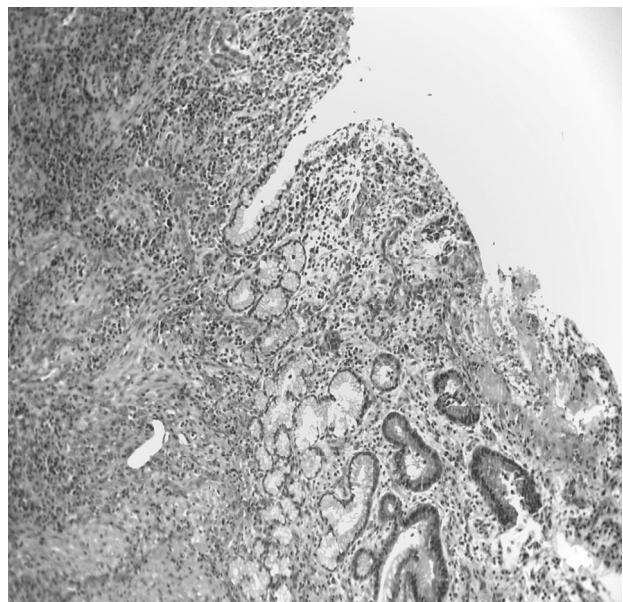


Figure 5 - Histological image of the gastric ulcer (hematoxylin-eosin x20).

Discussion

HH consists of herniation of elements of the abdominal cavity through the esophageal hiatus (usually stomach) of the diaphragm into mediastinum (1, 2, 9). The most comprehensive classification scheme recognizes two types of hiatal hernia (9). With type I, or sliding hiatal hernia, there is a widening of the muscular hiatal tunnel and circumferential laxity of the phreno-esophageal membrane, allowing a portion of the gastric cardia to herniate upward; with type II hernia the leading edge is the gastric cardia while with type III it is the gastric fundus (9). The esophago-gastric junction maintains its native position in the para-esophageal hernia while it is displaced upward with the sliding hernia (9). Para-esophageal HHs represent about 5-10% of all hiatal hernias, and are mainly represented in females (5, 9, 10]. While sliding HHs are frequently related to GERD, the paraesophageal ones are mainly asymptomatic and usually not associated to GERD (1, 11, 12). The most important symptoms are dyspepsia, hiccup and epigastric pain (maybe due to a transitory gastric twist) (12). Most important complications are dysphagia (related to esophageal compression), bleeding (secondary to ulcers or erosions) and twist (volvulus) (11-13). Usually incidental findings in about 5.2% of patients with HH performing EGDS, Cameron lesions represent gastric erosions and ulcers on mucosal folds in the distal neck of HHs (11, 14).

The pathogenesis of Cameron lesions is poorly understood and no univocal pathogenesis has been described since now: the causes ranging from mechanical trauma secondary to diaphragmatic contraction from respiratory excursions and acid injury, to ischemia (secondary to a vascular stasis) (7, 11, 15, 16).

It is more likely that the etiology could be multifactorial, including genetic and phenotypic tendency, patient risk factors such as co-morbidities and medication use (15). The prevalence is also related to the size of the HH with a 10-20% risk in hernias more than 5 cm (17).

Cameron lesions can be round or ellipsoid, even if linear forms are the most common ones (17, 18).

Cameron lesions, usually presenting as linear erosions along a mucosal fold of distal neck of HH, are

the milder form; Cameron ulcers, appearing macroscopically as ulcerations of the gastric wall, are the more serious presentation (19, 20).

Firstly described by Cameron and Higgins in 1986, these lesions, even if uncommon cause of severe upper-gastrointestinal bleeding, acquire relevance because of the possibility of causing chronic bleeding (with microcytic iron deficiency anemia) or acute and severe hemorrhage requiring blood transfusions, up to a severe hypovolemic shock (15, 21).

In 2001 Ruhl CE et al. published a study showing that patients with HH more than 5 cm in size had a significantly higher association of iron-deficiency anemia as compared to those with esophagitis (12).

More alarming are lesions presenting as severe, acute gastro-intestinal bleeding.

Diagnosis is established based upon anamnesis, clinical examination, barium-contrast X-rays and EGDS (with the opportunity to perform biopsies) (13, 14, 18-22).

In 2011 Chun CL et al. published an interesting study underscoring that Cameron lesions may be missed on endoscopic inspection if the clinician is not actively searching for them both due to lack of variable degrees of severity (ranging from small erosions to bigger ulcerations) and due to the relative rarity of these lesions (18). In their analysis the main conclusions are: first, findings such as edema, erythematous changes, and ecchymosis could suggest Cameron lesions in the gastric mucosal folds; second, a complete evaluation of the neck of the hernia, including antegrade, retrograde and perpendicular views, is important for the detection of Cameron lesions; third, identification of these lesions requires both a meticulous and knowledgeable clinician/endoscopist, suggesting that Cameron ulcers remain an important diagnostic challenge (18, 23).

Patients with Cameron lesions typically respond successfully to medical treatment consisting of iron supplementation with or without acid-suppression therapy (6-8).

In 1996 Weston et al. reported a rebleeding rate of 6.3% for patients treated medically (13).

Subsequent treatments are reserved to patients not responding to medical treatment and consist of endoscopic or surgical approaches (7, 24).

In 2013 Camus et al. published an important manuscript on Endoscopy, reporting the endoscopic

approach for hemostasis of Cameron ulcers; unfortunately, nowadays any guideline for appropriate techniques for endoscopic hemostasis have not yet been established (7). Nevertheless, in case of severe acute bleeding from Cameron ulcers, an efficient endoscopic approach may offer definitive hemostasis, reducing the risk of an invasive urgent surgical operation (24, 25).

Main endoscopic treatments consist of hemoclip positioning, thermal coagulation, endoscopic band ligation and hemostatic agents applying (such as cyanoacrylate or Hemospray®) (7, 8, 16, 25). Novel emerging endoscopic treatments for patients with GERD and small hiatal hernias, such as MUSE® system of EsophiX® device have not been evaluated for Cameron ulcer treatment yet (16).

As underlined in 2005 by Moschos J et al., surgery is required both in case of acute bleeding refractory to medical or endoscopic treatment and in case of severe sliding hernias with Cameron lesions (24). Moreover, as showed by Cougard P et al., long-term recurrence rates are extremely low following surgery (20).

Usually, elective surgical approach to Cameron ulcers consists of repairing the para-esophageal hernia, with or without fundoplication (7, 8, 24).

Unfortunately, few cases of surgery in patients with life-threatening bleeding from Cameron ulcers failing endoscopic therapy have been reported in literature since now.

In the present case the endoscopic treatment was unsuccessful with multiple episodes of melena, leading to an hemodynamic instability. After a multidisciplinary discussion, the patient was referred to surgery, putting the surgeons in front of a difficult choice: which kind of surgical approach perform? Literature shows a few cases like this, so that no univocal recommendations can be given.

Thus far, recently, Tan et al. published an article reporting the first case of a single-incision laparoscopic transgastric approach to secure an acute gastrointestinal hemorrhage (8). The conclusion of this paper is that in front of bleeding gastric ulcers not amenable to endoscopic or radiological interventions or whenever the patient becomes unstable due to the loss of blood, this surgical minimally invasive option could be taken into consideration to treat a bleeding ulcer (8). In this specific case the patient presented an uneventful recovery and subsequently

underwent a successful laparoscopic para-esophageal hernia repair.

In our case, considering the several comorbidities of the patient and the life-threatening bleeding from the Cameron ulcer, the surgeons decided to treat in the same operation both the para-esophageal hernia and the active bleeding ulcer. In the operative theatre the patient was in a supine position with both arms out. A pneumo-peritoneum with carbon dioxide was established at a pressure of 12 mmHg with high flow. Inotropic requirements and ventilatory settings remained unchanged before and after the establishment of pneumo-peritoneum.

After opening esophago-gastric lamina, the presence of the para-esophageal hernia was confirmed. Once reduced the stomach into the peritoneal cavity, an intra-operative EGDS confirmed the presence of a large type Forrest 1A ulcer on a mucosal fold of the neck of the HH. In the operative theatre, in order to ensure a complete resection of the gastric wall involved by the ulcer, the surgeons decided to perform an endoscopically-guided atypical and since now undescribed gastric resection with a mechanical stapling positioned along the fundic gastric wall. A careful hemostasis control was done, and a Blake® drainage 19 French in diameter size was placed along the suture. The surgical operation was totally conducted in laparoscopy and the total operative time was about 100 minutes. Estimated blood loss was less than 400 ml.

Unfortunately, nowadays few cases of surgical treatment of life-threatening bleeding for Cameron ulcers have been described, therefore no univocal and standardized recommendation exists (7, 8, 11, 25). As a result of this lack of knowledge, treatment of severe bleeding due to Cameron ulcer remains very challenging, and the acquisition of further consistent data is needed.

Conclusions

Cameron ulcers remain a rare cause of upper gastrointestinal bleeding (26, 27). After an overview of the current literature, the Authors recommend medical and endoscopic treatment first, reserving surgery for patients who fail medical/endoscopic therapy or whenever an hemodynamic instability occurs.

Nowadays, few cases of urgent surgical approach-

es for severe bleeding related to Cameron ulcers have been described, so that the decision of the best surgical treatment remains a very challenging problem.

No univocal indication exists about the surgical treatment of life-threatening hemorrhage due to Cameron ulcers, so that, actually, the most important challenge of surgical treatment lies in choosing a tailored strategy for each patient, taking into account age, general health condition, comorbidities, presence of hemodynamic instability and time since diagnosis (8).

We think that, out of an emergency situation, the patients should be addressed to hospitals at high volume of gastric procedures, in order to assess the best standard of care.

Abbreviations

GERD: gastro-esophageal reflux disease.

EGDS: esophagogastroduodenoscopy.

PPI: proton-pump inhibitor.

HH: hiatal hernia.

CT: computed tomography.

Ethical approval

Not requested.

Funding

None funding.

References

1. Hyun JJ, Bak YT. Clinical significance of hiatal hernia. *Gut Liver*. 2011 Sep;5(3):267-77.
2. Yakut M, Kabaçam G, Öztürk A, Soykan I. Clinical characteristics and evaluation of patients with large hiatal hernia and Cameron lesions. *South Med J*. 2011 Mar;104(3):179-84.
3. Kahrilas PJ, Kim HC, Pandolfino JE. Approaches to the Diagnosis and Grading of Hiatal Hernia. *Best Pract Res Clin Gastroenterol*. 2008;22(4):601-16.
4. Oleynikov D, Jolley JM. Paraesophageal hernia. *Surg Clin North Am*. 2015 Jun;95(3):555-65.
5. Schieman C, Grondin SC. Paraesophageal hernia: clinical presentation, evaluation and management controversies. *Thorac Surg Clin*. 2009;19:473-84.
6. Gray DM, Kushnir V, Kalra G, Rosenstock A, Alsakka MA, Patel A, Sayuk G, Gyawali CP. Cameron lesions in patients with hiatal hernias: prevalence, presentation, and treatment outcome. *Dis Esophagus*. 2015 Jul;28(5):448-52.
7. Camus M, Jensen DM, Ohning GV, Kovacs TO, Ghassemi KA, Jutabha R, Machicado GA, Dulai GS, Hines OJ. Severe upper gastrointestinal hemorrhage from linear gastric ulcers in largehiatal hernias: a large prospective case series of Cameron ulcers. *Endoscopy*. 2013;45(5):397-400.
8. Tan CHN, Kim G, So J, Shabbir A. Single-Incision Laparoscopic Transgastric Underrunning and Closure of Cameron Ulcers in Acute Gastrointestinal Bleeding. *J Gastrointest Surg*. 2018 Mar;22(3):553-6.
9. Yu HX, Han CS, Xue JR, Han ZF, Xin H. Esophageal hiatal hernia: risk, diagnosis and management. *Expert Rev Gastroenterol Hepatol*. 2018 Apr;12(4):319-29.
10. Draaisma WA, Gooszen HG, Tournioji E, Broeders IA. Controversies in paraesophageal hernia repair: a review of literature. *Surg Endosc*. 2005;19:1300-8.
11. Kapadia S, Jagroop S, Kumar A. Cameron ulcers: an atypical source for a massive upper gastrointestinal bleed. *World J Gastroenterol*. 2012 Sep 21;18(35):4959-61.
12. Ruhl CE, Everhart JE. Relationship of iron-deficiency anemia

Authors' contribution

Damiano Bisogni, Andrea Valeri: substantial contributions to conception and design, writing the study, drafting the article, final approval of the version to be published.

Luca Talamucci, Roberto Manetti, Alessio Biagio Filippo Giordano, Massimiliano Ardu: bibliography collection, drafting the article, final approval of the version to be published.

Riccardo Naspetti, Paolo Prosperi: bibliography collection, drafting the article, final approval of the version to be published.

Acknowledgements

The Authors would like to acknowledge Michele Rossi, Anna Maria Di Bella, Veronica Iacopini, Giovanni Alemanno and Carlo Bergamini (Medical Doctors) for assistance in creating the work.

Conflicts of interest

None.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Guarantor of submission

The corresponding Author is the guarantor of submission.

- with esophagitis and hiatal hernia: hospital findings from a prospective, population-based study. *Am J Gastroenterol*. 2001 Feb;96(2):322-6.
13. Weston AP. Hiatal hernia with cameron ulcers and erosions. *Gastrointest Endosc Clin N Am*. 1996;6:671-9.
 14. Cameron AJ, Higgins JA. Linear gastric erosion. A lesion associated with large diaphragmatic hernia and chronic blood loss anemia. *Gastroenterology*. 1986;91:338-42.
 15. Gupta P, Suryadevara M, Das A, Falterman J. Cameron Ulcer Causing Severe Anemia in a Patient with Diaphragmatic Hernia. *Am J Case Rep*. 2015 Oct 15;16:733-6. PubMed PMID: 26467083; PubMed Central PMCID: PMC4612144.
 16. Moskovitz M, Fadden R, Min T, et al. Large hiatal hernias, anemia, and linear gastric erosion: Studies of etiology and medical therapy. *Am J Gastroenterol*. 1992;87:622-6.
 17. Nguyen N, Tam W, Kimber R, Roberts-Thomson IC. Gastrointestinal: Cameron's erosions. *J Gastroenterol Hepatol*. 2002;17:343.
 18. Chun CL, Conti CA, Triadafilopoulos G. Cameron ulcers: you will find only what you seek. *Dig Dis Sci*. 2011 Dec;56(12): 3450-2.
 19. Appleyard MN, Swain CP. Endoscopic difficulties in the diagnosis of upper gastrointestinal bleeding. *World J Gastroenterol*. 2001;7:308-12.
 20. Cougard P, Sala JJ, Favre JP, Viard H. Ulcers of the neck of hiatal hernias. 15 cases. *J Chir (Paris)*. 1985 Jun-Jul;122(6-7):399-402.
 21. Maganty K, Smith RL. Cameron lesions: Unusual cause of gastrointestinal bleeding and anemia. *Digestion*. 2008;77(3):214-8.
 22. Aypak C, Çakmak N, Görpelioglu S. An unusual cause of anemia: Cameron ulcer. *Cukurova Med J*. 2013;38:315-18.
 23. Zaman A, Katon RM. Push enteroscopy for obscure gastrointestinal bleeding yields a high incidence of proximal lesions within reach of a standard endoscope. *Gastrointest Endosc*. 1998;47:372-6.
 24. Moschos J, Pilpilidis I, Kadis S, Antonopoulos Z, Paikos D, Tzilves D, Katsos I, Tarpagos A. Cameron lesion and its laparoscopic management. *Indian J Gastroenterol*. 2005 Jul-Aug;24(4):163.
 25. Lin CC, Chen TH, Ho WC, et al. Endoscopic treatment of a Cameron lesion presenting as lifethreatening gastrointestinal hemorrhage. *J Clin Gastroenterol*. 2001;33:423-4.
 26. Cameron AJ. Incidence of iron deficiency anemia in patients with large diaphragmatic hernia. A controlled study. *Mayo Clin Proc*. 1976;51:767-9.
 27. Gallego-Pérez B, Martínez-Crespo JJ, Marín-Bernabé CM, Gajownik U. Cameron ulcers: two clinical presentations of an unusual upper gastrointestinal hemorrhage. *Gastroenterol Hepatol*. 2014 Dec;37(10):594-5.