Valentino’s Syndrome (with Retroperitoneal Ulcer Perforation): A Rare Clinico-Anatomical Entity

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Patient: Male, 51-year-old
Final Diagnosis: Valentino syndrome • perforated duodenal ulcer to the retroperitoneum
Symptoms: Epigastric pain • loss of appetite • nausea • right lower quadrant abdominal pain
Medication:
Clinical Procedure: Exploratory laparotomy
Specialty: Surgery

Objective: Rare disease
Background: In the emergency department pain in the right lower quadrant is a frequent finding and is related to a wide variety of diseases, the most common of which is acute appendicitis. An unusual presentation of pain in the right iliac fossa is due to perforation of a duodenal peptic ulcer. The fluid that originates from the perforated ulcer moves through the paracolic sulcus to the right iliac fossa and causes irritation of the peritoneum and even chemical peri-appendicitis, thereby imitating all the usual causes of pain in the right lower quadrant. This condition is known as Valentino’s syndrome, named after the Italian actor Rudolph Valentino.

Case Report: The aim of this case report was to review the current published literature regarding Valentino’s syndrome and report on a case involving a 51-year-old male who was admitted to our surgical department with right lower quadrant pain and suspicion of acute appendicitis. An exploratory laparotomy was performed, from which a retroperitoneal perforation of a duodenal ulcer was found; suture closure was then applied. The patient’s post-operative course was uncomplicated, and he was discharged 9 days after the operation.

Conclusions: Surgeons should be alert for this rare condition imitating acute appendicitis, and the differential diagnosis of right lower quadrant pain should include peptic ulcer perforation.

MeSH Keywords: Abdomen, Acute • Appendicitis • Peptic Ulcer

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/922647
Background

Surgeons in the emergency department often encounter patients with pain located in the right lower quadrant. It is a challenging condition to diagnosis because many conditions can cause this symptom. The differential diagnosis of acute right lower quadrant pain is more complicated in women than it is in men, because acute gynecological conditions can be misdiagnosed as acute appendicitis. The main target of a surgeon is to exclude an acute surgical abdomen that needs immediate surgical treatment. One of the most common causes of pain in the right iliac fossa is acute appendicitis, presenting with symptoms like fever, loss of appetite, nausea, or vomiting. Other conditions that can cause right lower quadrant pain include ureteric colic, diverticulitis, or rupture of a diverticulum, mucocoele of appendix, perforated cholecystitis, pancreatitis, colitis and more. Additionally, in women, acute gynecological conditions that can mimic acute appendicitis include ovarian torsion, ruptured ectopic pregnancy, endometriosis, infarcted uterine leiomyoma, and pelvic inflammatory diseases [1–5]. Although Valentino’s syndrome is rare, it remains one of the causes of acute right lower quadrant pain, and it should always be included in the differential diagnosis [6]. Rudolph Valentino was hospitalized in the New York Polyclinic Hospital in 1926, where an appendectomy was performed, but he later died due to pleurisy.

A duodenal ulcer with perforation through the retroperitoneum leads to leakage of gastric contents in the right iliac fossa, resulting in the main symptom aforementioned and mimicking acute appendicitis. Early diagnosis is essential since if there is a delay in the surgical treatment of a perforated peptic ulcer then morbidity and mortality are increased [1,2]. We found only a few reported cases of Valentino’s syndrome in the existing literature. Usually, patients undergo an operation for an acute surgical abdomen with suspicion of acute appendicitis, but when the appendix is found to be normal without inflammation, intraoperative investigation leads to diagnosis of a perforated peptic ulcer.

Case Report

A 51-year-old Caucasian male presented to the emergency department with persistent abdominal pain, nausea, and loss of appetite. He mentioned that symptoms started approximately 6 hours before visiting the emergency department. Initially, the patient suffered from an epigastric pain of acute onset that radiated to the right iliac fossa. He described the pain to be stabbing in character, accompanied by 3 episodes of vomiting. He did not mention fever, rigor, or any other symptoms. The patient had a history of duodenal ulcer diagnosed 10 years ago during an endoscopic examination because he experienced a burning sensation in the stomach and abdominal pain after eating. Tests were positive for Helicobacter pylori and the patient was treated with antibiotics and proton pump inhibitors. He was not taking any medications, and he was not abusing alcohol, tobacco, or any other substances. He had a surgical removal of his palatine tonsils as a child. During the examination, he was tachycardic with increased respiratory rate. Percussion of the patient’s abdomen revealed pain located both in the epigastrium and the right lower quadrant. During percussion of the left iliac fossa the pain migrated to the right lower quadrant. In addition, palpation of the abdominal wall showed tenderness and guarding. All clinical signs of acute appendicitis and peritonitis such as McBurney, Rovsing, Blumberg, psoas and obturator were positive. His Alvarado score was 8 out of 10.

Laboratory investigations revealed neutrophilic leukocytosis, with a white blood cell count of 17 700 cells/mL of blood and 86.5% neutrophils. C-reactive protein (CRP) level was 0.332 mg/dL, within normal value. The remaining blood and urine tests were normal. There were no pathological findings on chest and abdominal x-rays (Figure 1). His electrocardiogram (ECG) showed sinus tachycardia.

Prior to his admission to the surgical department, we intervened with venous and Foley catheters and a Levin naso-gastric tube. Initial antibiotic treatment employed sultamicillin, which was later changed to tazobactam/piperacillin. The patient was transferred to the operating theater on the same day, 2 hours after his admission to the surgical department, and an exploratory laparotomy was performed. In our surgical department we perform exploratory laparotomies for all acute surgical abdomens of unknown origin and despite the fact that acute appendicitis seemed to be the most likely diagnosis, the patient’s history of duodenal ulcer created doubts. During the operation, the appendix had no signs of inflammation and free intraperitoneal fluid was found in the right paracolic gutter and iliac fossa. An extensive Kocher maneuver was performed and a perforation of a duodenal ulcer to the retroperitoneum was found along with a small amount of fluid in the retroperitoneal cavity. Intermittent closure with monofilament absorbable size 3-0 sutures was applied to the site of the leakage without omentoplasty and 2 corrugated drains were placed (Figure 2). The first drain was placed in the area of the perforation and the second in the right paracolic gutter. Therapy with proton pump inhibitors was initiated, and the antibiotic treatment with tazobactam/piperacillin was continued. The patient’s postoperative course was uneventful, and he was discharged in good overall health 9 days after the operation.

Discussion

Peptic ulcers are open sores that develop on the inside of the stomach and the duodenum and are divided into gastric and
Figure 1. (A) Chest (posterior-anterior view); (B) abdominal x-rays (erect position) demonstrating the absence of free intraperitoneal air.

Figure 2. Intraoperative findings. (A) After extensive Kocher maneuver, we demonstrate the presence of the perforation at the superior part of the duodenum. (B) The application of sutures (type: monofilament absorbable, size: 3-0) at the site of the leakage.

duodenal types. Infection with *Helicobacter pylori*, and long-term use of nonsteroidal anti-inflammatory drugs (NSAIDs) and aspirin are the most common causes of peptic ulcer disease. Uncomplicated peptic ulcer disease presents with symptoms like epigastric pain, bloating, heartburn, nausea, and a burning stomach sensation. Less often, ulcers can cause severe symptoms such as melena and hematemesis [7]. Perforation of these ulcers is considered a surgical emergency with increased morbidity and mortality if the duration of the perforation exceeds 24 hours and the size is greater than 1 cm.
Perforated peptic ulcers usually present with a sharp epigastric pain of sudden onset that sometimes radiates to the shoulder, indicating free abdominal air. The anterior duodenal wall is the most common site of perforation, accounting for 60% of the cases, and it is the most frequent type in Western countries [8]. The perforation of a duodenal ulcer through the retroperitoneum, presenting with pain in the right lower quadrant because of fluid leakage and accumulation in the right paracolic gutter, is known as Valentino’s syndrome. It presents as an acute surgical abdomen mainly mimicking acute appendicitis, and it leads the patient straight to the surgical theater; thus, the final diagnosis is established during the operation [1–3,9,10]. After extensive literature review, we found only 5 cases, and in half the cases, the diagnosis was formed prior to operation with a computed tomography (CT) scan, which is the examination of choice. Signs found on CT imaging include right retroperitoneal free air, mainly around the right kidney, known as “veiled right kidney sign”, thickening of the wall of the duodenum, and free air around the duodenum. Simple chest and abdominal x-rays may illustrate free intraperitoneal air under the right diaphragm or around the right kidney, confirming the hypothesis for perforation of the duodenum [2,3,11]. Inability to perform a CT scan at this point suggests that an exploratory laparoscopy should be performed, which is recommended for the investigation of an acute surgical abdomen when there is a suspicion, but not a definite diagnosis of acute appendicitis [12].

In the current case report, the patient had no signs of free intraperitoneal air on x-ray images; he had an acute abdomen with a strong suspicion of acute appendicitis and there was no ability to arrange a CT scan as the CT scanner unexpectedly went out of order and was unavailable, so an exploratory laparotomy was performed. Surgeons should suspect peptic ulcer perforation in a patient with signs similar to acute appendicitis and a history of duodenal ulcer. There are a few options regarding the management of a perforated peptic ulcer. The Taylor method is non-invasive, and it is based on combination of antibiotic treatment, \textit{H. pylori} triple therapy, intravenous fluids, naso-gastric tube aspiration and clinical observation of the patient. When surgical operation is preferred and the site of perforation is identified, then simple closure may be performed. Surgical techniques that can be used are simple closure with intermittent sutures, with or without omentoplasty; an omental plug drawn into the site of perforation and sutured over; and finally, an omental patch after Graham technique. Simple closure during exploratory laparoscopy is the minimally invasive technique, followed by eradication of \textit{H. pylori} [8,13,14].

Conclusions

Perforation of a duodenal ulcer through the retroperitoneum rarely may present by mimicking acute appendicitis. Surgeons should include perforation of a peptic ulcer in the differential diagnosis of patients presenting with right lower quadrant pain, and a CT scan should be the examination of choice. This report adds to the published literature on cases of Valentino’s syndrome. This case was diagnosed during operation as there was a strong suspicion of acute appendicitis and no ability to perform a CT scan. Surgeons should be aware of this syndrome, and if in our case, a CT scan had been performed prior to surgery, maybe the treatment would have been different.

Conflict of interest

None.

References:

1. Wijegoonewardene S, Stein J, Cooke D, Tien A: Valentino’s syndrome a perforated peptic ulcer mimicking acute appendicitis. BMJ Case Rep, 2012 ;2012: bcr0320126015
2. Ishi A, Omogbose S, Osime O: Surgeons beware: It may not be acute appendicitis. Arch Int Surg. 2017; 7: 139–41
3. Mohan CP, Kabalimurthy J, Balamurugan E, Jayavarmaa R: A rare case of Valentino’s syndrome. Int Surg J. 2018; 5: 2933–35
4. Dural R, Hoque H, Ng P: The acute abdomen – commonly missed and misdiagnosed conditions. Review: Webmed Central Surgery, 2010; 1: WMC001036
5. Patel NB, Wenzke DR: Evaluating the patient with right lower quadrant pain. Radiol Clin North Am, 2015; 53: 1159–70
6. Mbarushimana S, Morris-Stiff G, Thomas G: Atypical presentation of perforated peptic ulcer disease in a 12-year-old boy. BMJ Case Rep, 2014; 27: bcr2014204716
7. Malfertheiner P, Chan FK, McColl KE: Peptic ulcer disease. Lancet, 2009; 374: 1449–61
8. Bertleff MJ, Lange JF: Perforated peptic ulcer disease: A review of history and treatment. Dig Surg, 2010; 27: 161–69
9. Amann C, Austin A, Rudinsky S: Valentino’s syndrome: A life-threatening mimic of acute appendicitis. Clin Pract Cases Emerg Med, 2017; 1: 44–46
10. Ramirez-Ramirez M, Villanueva-Saenz E: Valentino’s syndrome. Perforated peptic ulcer with unusual clinical presentation. Rev Gastroenterol Mex, 2016; 81: 225–26
11. Mahajan P, Abdalla M, Purayil N: First report of preoperative imaging diagnosis of a surgically confirmed case of Valentino’s syndrome. J Clin Imaging Sci, 2014; 4: 28
12. Golash V, Willson PD: Early laparoscopy as a routine procedure in the management of acute abdominal pain: A review of 1,320 patients. Surg Endosc, 2005; 19: 882–85
13. Ramírez SA, Devlin HB: Perforated duodenal ulcer. Br J Surg, 1987; 74: 81–82
14. Tomitchong P, Siribumrongwong B, Vilaichone RK et al: Systematic review and meta-analysis: \textit{Helicobacter pylori} eradication therapy after simple closure of perforated duodenal ulcer. Helicobacter, 2012; 17: 148–52

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