Paraduodenal hernias are the most common type of congenital internal hernia. Because of its overall rare incidence, this entity is often overlooked during initial assessment of the patient. Lack of specific diagnostic criteria also makes diagnosis exceedingly difficult, and the resulting diagnostic delays can lead to tragic outcomes for patients. Despite these perceived barriers to timely diagnosis, there may be specific radiographic findings that, when combined with the appropriate constellation of clinical symptoms, would aid in diagnosis. This patient first presented at 8 years of age with vague symptoms of postprandial emesis, chronic abdominal pain, nausea, and syncope. Over the span of 6 years he was evaluated 2 to 3 times a year with similar complaints, all of which quickly resolved spontaneously. He underwent multiple laboratory, imaging, and endoscopic studies, which were nondiagnostic. It was not until he developed signs of a high-grade obstruction and extremis that he was found to have a large left paraduodenal hernia that had volvulized around the superior mesenteric axis. This resulted in the loss of the entire superior mesenteric axis distribution of the small and large intestine and necrosis of the duodenum. In cases of chronic intermittent obstruction without clear etiology, careful attention and consideration should be given to the constellation of symptoms, imaging studies, and potential use of diagnostic laparoscopy. Increased vigilance by primary care and consulting physicians is necessary to detect this rare but readily correctable condition.

Internal hernias (IHs) are a rare clinical entity that cause 0.6% to 5.8% of small bowel obstructions.1 There are 2 broad categories of IH: acquired and congenital. These are further categorized by location, which include paraduodenal, transmesenteric, and supra- or perivesical IHs and those located at the Foramen of Winslow.2,3 Paraduodenal hernias (PDHs) are thought to be the most common type of congenital IH.2,3 However, with the increasing incidence of procedures that require Roux-en-Y reconstruction, such as gastric bypass surgery, Kasai, choledochal cyst excision, and pediatric liver transplantation, the incidence of acquired IH is increasing.4 Left PDH refers to a herniation of the small bowel into a mesenteric defect to the left of the inferior mesenteric vein (IMV) with a hernia sac within the leaflets of the left colon mesentery.5 Symptoms can range from intermittent partial bowel obstruction in cases of spontaneously reducing IH to acute high-grade obstruction that occurs with incarceration or volvulus.6 Radiographic diagnosis is made difficult because of the often intermittent nature of the disease, in which spontaneous reduction in and resolution of symptoms can occur by the time diagnostic imaging is performed.2,7 Even when present,
radiographic signs are not always readily apparent.7 Without appropriate awareness, many IHs may be undiagnosed or diagnosed incorrectly, resulting in serious consequences for the patient.

CASE REPORT

A 14-year-old boy with attention-deficit/hyperactivity disorder, constipation, and chronic intermittent abdominal pain presented to the emergency department with acute-onset severe abdominal pain and vomiting. Over the past 6 years, the patient reported having similar episodes of abdominal pain, vomiting, and syncope. During this interval he was seen in the emergency department 10 to 15 times between 2 institutions; surgical consultation was requested at one of these visits. During each encounter his symptoms resolved spontaneously, all diagnostic workups were negative, and the patient was discharged from the hospital without a definitive diagnosis. Further confounding the presentation was a complex psychosocial environment that included a parental death. In addition to the most common etiologies for abdominal pain, he was also evaluated for sickle cell disease and acute intermittent porphyria. Kidney, ureter, and bladder radiographs (KUB), abdominal and pelvic computed tomography (CT) scans, electrocardiogram, echocardiogram, esophagastroduodenoscopy, and colonoscopy were all nondiagnostic.

Ten days before admission the patient was seen in the ED and discharged with a presumed psychological etiology of abdominal pain. At gastroenterology outpatient evaluation 3 days later his pain was attributed to possible autonomic nervous system dysfunction caused by his attention-deficit/hyperactivity disorder medication, which was discontinued, and he was referred to psychiatry. Three days before admission he was admitted to an outside institution where he was diagnosed with abdominal migraines, treated with valproic acid with resolution of symptoms, and then discharged after 24 hours.

On the day of admission, because of lack of a clear diagnosis, referral to psychology and neurology was considered for evaluation of possible abdominal migraine, conversion disorder, or malingering. The morning of admission, the patient became highly agitated and altered in the ED. He was admitted to the pediatrics service after being given valproic acid and diphenhydramine. Repeat KUB at this time was unremarkable. On the evening of admission he was found to be anuric, increasingly altered, hypotensive, tachycardic, and tachypneic. Multiple laboratory derangements were noted. He was moved to the ICU where he had an episode of frank hematemesis. At this point, a surgical consultation was ordered. Repeat KUB showed a small bowel obstruction and possible pneumoperitoneum. The patient was then taken to the operating room for an emergent exploratory laparotomy.

Upon entering the patient’s abdomen, the surgical team encountered grossly necrotic bowel (Fig 1A). Further inspection revealed a large left PDH defect (Fig 1B), which contained the midgut, cecum, appendix, and a portion of the right colon. The herniated bowel was also found to be volvulized about the axis of the superior mesenteric artery. Over the course of several operations, the patient’s bowel from the second portion of the duodenum to the descending colon was resected. He ultimately underwent 12 operations and spent 3 weeks in the ICU. His postoperative course was complicated by central catheter–associated sepsis and renal failure requiring dialysis. He also suffered a duodenal stump leak, which was controlled by using multiple intraabdominal drains, which remained in place for 1 year. He is currently under evaluation for a multivisceral transplant and is being maintained on full parenteral nutrition.

Retrospective review of the patient’s CT scan revealed the presence of a “sac-like” mass of bowel loops with associated engorged vessels (Fig 2), as well as an IMV, which was anteriorly displaced (Fig 3).

DISCUSSION

The patient in this case had intermittent abdominal discomfort for 6 years. Coupled with his underlying social stressors, the effects of multiple hospital encounters and chronic pain resulted in significant emotional distress, nutritional deficit, and missed school. Unfortunately, delayed diagnosis is all too common in PDH, and patients often remain undiagnosed until signs of high-grade obstruction or bowel compromise.
When strangulation is present, mortality can approach 50%. This makes expedient diagnosis and intervention paramount.

Clinical diagnosis of PDH is challenging due to the lack of discrete signs and symptoms. Abdominal examinations in this case were never indicative of serious underlying pathology until the patient became acutely ill. Before the occurrence of incarceration or strangulation, distention is rarely present and symptoms are often vague and can last years to decades, especially in cases of PDH compared with other types of IH. Symptoms range from mild dyspepsia and nonlocalized abdominal pain to nausea and emesis. Postprandial abdominal pain and changes in pain based on body position have also been reported. These symptoms often resolve with spontaneous reduction of the hernia. Therefore, diagnosis generally must be made with appropriate clinical suspicion and fortuitously timed radiographic imaging. Elective diagnostic laparoscopy could also have aided in obtaining a diagnosis in this patient.

Before the advent of CT, radiographic diagnosis of IH was made by using upper gastrointestinal series. Left PDH typically presents as sac-like small bowel loops left of the duodenum with possible mass effect on the transverse colon or posterior stomach. Recently, multidetector row CT has become the diagnostic test of choice for PHDs because it allows increasingly accurate visualization of the bowel mesentery and its associated vasculature. It has several other significant advantages over traditional upper gastrointestinal series including that it is prevalent, noninvasive, and can be performed quickly, ideally while the patient is symptomatic.

Most of the literature related to CT diagnosis of left PDH consists of isolated case reports or small series. A review of 54 cases of IH identified 6 classified as PDH, 2 of which had CT images available for review. The presence of a sac-like cluster of small bowel loops located between the pancreas and stomach, mild small bowel dilation, and closely approximated engorged mesenteric vessels was consistent with PDH. Mass effect on the posterior stomach, duodenjejunal junction, and
transverse colon can also be present.\textsuperscript{6} The presence of the inferior mesenteric vessels at the anterior neck of the hernia sac have been described in previous angiographic studies.\textsuperscript{9} An association between the displacement of these vessels and left PDH has also been noted with the use of CT imaging.\textsuperscript{5,15,16} The most consistently reported signs in the literature that suggest the presence of a PDH are as follows: the presence of bowel between the stomach and pancreas or the transverse colon and left adrenal gland, displacement of the IMV upward and anteriorly, and presence of an encapsulated bowel sac lateral to the duodenum.\textsuperscript{5,7,9–11,13,15,17–19}

Despite the presence of well-reported diagnostic criteria, many nonobstructive IHs go undiagnosed or misdiagnosed.\textsuperscript{5,20} It is important to note that these radiographic findings can be subtle, and a lack of awareness likely contributes to missed diagnoses. In a small sample, when radiologists were asked to specifically evaluate for a “sac-like” mass of small bowel, mass effect on the posterior wall of the stomach, and displacement of the IMV, the presence of PDH was able to be identified accurately on CT.\textsuperscript{17} In addition, retrospective review of this patient’s abdominal CT performed 16 months before diagnosis revealed subtle rostral and leftward displacement of the IMV, engorged mesenteric vessels, and a sac-like mass of small bowel loops. Perhaps with the appropriate clinical suspicion, these insidious clinical entities may be elucidated with imaging.

Given the potential for deleterious outcomes, the presence of an IH should be aggressively ruled out in patients with long-standing symptoms of chronic intermittent bowel obstruction, especially in patients without a history of significant abdominal trauma or abdominal surgery when more obvious and prevalent pathologies have been excluded. An extensive workup may not be necessary and may only result in higher costs. In this patient, comprehensive imaging and invasive diagnostic procedures did not contribute toward the diagnosis. Previous radiographic images should also be reviewed for subtle signs that may have been unnoticed. Special attention should be given to the presence of small bowel adjacent to the stomach, sac-like bowel on CT, and IMV displacement. Patients may benefit from earlier surgical consultation and diagnostic laparoscopy to provide a potential definitive diagnosis and therapeutic intervention before the onset of bowel compromise. The presence of third-party records, especially imaging, should be aggressively solicited. Patients with long-standing undiagnosed symptoms may seek care at multiple institutions, making a complete clinical picture difficult to obtain. IH is a rare clinical entity with potentially catastrophic consequences for the patient, which may be prevented by awareness of the associated constellation of symptoms and radiographic signs. Clinical suspicion by care providers, appropriately communicated to radiology, is key to arriving at a timely and correct diagnosis.

ACKNOWLEDGMENT

We would like to acknowledge Ana Rodriguez PhD, for her editorial support.

REFERENCES


Congenital Left Paraduodenal Hernia Causing Chronic Abdominal Pain and Abdominal Catastrophe

Yan Shi, Amy E. Felsted, Prakash M. Masand, Brent A. Mothner, Jed G. Nuchtern, J. Ruben Rodriguez and Sanjeev A. Vasudevan

*Pediatrics*; originally published online March 23, 2015;
DOI: 10.1542/peds.2014-3701
Congenital Left Paraduodenal Hernia Causing Chronic Abdominal Pain and Abdominal Catastrophe
Yan Shi, Amy E. Felsted, Prakash M. Masand, Brent A. Mothner, Jed G. Nuchtern, J. Ruben Rodriguez and Sanjeev A. Vasudevan

*Pediatrics*; originally published online March 23, 2015;
DOI: 10.1542/peds.2014-3701

The online version of this article, along with updated information and services, is located on the World Wide Web at:
/content/early/2015/03/17/peds.2014-3701