A rare clinical presentation of heterotopic gastric mucosa of the jejunum: A case report and review of the literature

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Article info

Article history:
Received 25 May 2014
Received in revised form 22 June 2014
Accepted 27 June 2014

Key words:
Heterotopic gastric mucosa
Chronic abdominal pain
Failure to thrive

Abstract

Heterotopic gastric mucosa (HGM) of the small bowel is a congenital disorder with a variable clinical presentation. Reported manifestations are gastrointestinal bleeding, intestinal obstruction or perforation, penetration into adjacent organs, and fistulization. Rarely, failure to thrive (FTT) is the clinical manifestation of HGM. We present the case of a baby girl with FTT due to chronic abdominal pain associated with recurrent episodes of abdominal distention, vomiting, and diarrhea. The cause was found to be HGM in the jejunum. The purpose of this paper is to describe this unusual clinical presentation of jejunal HGM.

Chronic abdominal pain and food intolerance in a child are common symptoms in many medical and surgical conditions, including infectious, allergic, inflammatory, neoplastic, mechanical, and motility disorders. We present the case of a baby girl with frequent episodes of abdominal pain, abdominal distention, vomiting, and diarrhea that resulted in failure to thrive (FTT). The cause of these symptoms was HGM in the jejunum.

1. Case report

A one-year-old girl presented with episodes of irritability, abdominal distention, vomiting, and diarrhea. Her past medical and family histories were unremarkable. The episodes lasted from a few days to weeks and required frequent admissions to a tertiary care hospital; an extensive workup did not reveal a definitive diagnosis. Infectious, allergic, immune, inflammatory, neoplastic, and mechanical obstructive causes were ruled out. The diagnosis of exclusion was chronic intestinal pseudo-obstruction syndrome (CIPO). During the episodes, the patient was treated with bowel rest and total parenteral nutrition. Oral elemental formula was tolerated between the episodes. The only medication prescribed was metoclopramide. At the age of four years the patient was referred to our institution for a second opinion.

At our hospital, laboratory tests completed included complete blood count, serum electrolytes including calcium, magnesium and phosphate, serum glucose, creatinine, liver transaminases, coagulation profiles, sickle cell screen, reticuloocyte count, zinc protoporphyrin, serum proteins, erythrocyte sedimentation rate (ESR), and antinuclear and anti-smooth-muscle antibody screen. All results were within normal limits except for a mild anemia, low albumin, and slightly elevated ESR. An infectious workup was negative including blood, urine, and stool cultures including for ova and parasites. A urinalysis was unremarkable and stool for occult blood was negative. An abdominal ultrasound did not reveal any abnormalities. An upper gastrointestinal (GI) contrast study with a small bowel follow through showed a dilated hypomotile segment of proximal jejunum followed by a short narrowed segment consistent with episodes of transient spasm. In both segments, there was marked mucosal irregularity and ulceration (Fig. 1). A computerized tomography (CT) scan of the abdomen with IV and oral contrast confirmed the findings from the upper GI contrast study and revealed several enlarged mesenteric lymph nodes (Fig. 2). Upper and lower GI endoscopy with multiple biopsies from the esophagus, stomach, duodenum, colon, and rectum revealed no pathological abnormalities.

A multidisciplinary meeting was conducted, including members from the pediatric specialties of gastroenterology, infectious...
disease, immunology, hematology-oncology, radiology and surgery. The conclusion was to proceed with surgical exploration, because of the lack of a definitive diagnosis and the possibility of bowel lymphoma. A laparoscopic approach was attempted but aborted due to intra-abdominal adhesions. Laparotomy revealed a dilated segment of proximal jejunum of 38 cm length and several enlarged mesenteric lymph nodes. The dilated segment was followed by an 8-cm narrowed segment with thickened walls (Fig. 3). The dilated proximal segment, distal thickened segment and the enlarged mesenteric lymph nodes were resected en-bloc (Fig. 4). Bowel continuity was restored with an end-to-end jejuno-jejunal anastomosis.

Tissue studies of the resected dilated segment revealed normal small bowel mucosa with multiple areas of HGM (Figs. 5 and 6). Multiple small ulcers were seen in the dilated segment. In the narrowed segment there was normal small bowel mucosa, no gastric mucosa, but multiple ulcers and significant scarring. Both ends of the resected jejunal segment contained normal intestinal mucosa and were free of HGM. Mesenteric lymph nodes showed reactive follicular hyperplasia.

Following the pathology report, a $^{99m}$Tc pertechnetate scan was performed to identify possible residual HGM in the abdomen and pelvis; the scan did not show any abnormal uptake.

Postoperatively, the patient was followed for three years. She is free of symptoms, tolerates a regular diet, and has reached acceptable growth parameters.

2. Discussion

Our patient’s initial presentation was very suggestive of an infectious process, the most common culprit in our community. An extensive initial workup did not reveal any infectious, allergic, immunologic, inflammatory, neoplastic, or mechanical causes. CIPO was the diagnosis of exclusion and thus the patient was managed at an outside institution for 3 years. However, she suffered from ongoing episodes and progressive FTT. At our hospital, repeated radiological assessments revealed the process to be localized to a segment of proximal jejunum consistent with a segmental bowel dilatation. Mucosal irregularity noted on the contrast GI study in the involved jejunal segment was interpreted as mucosal changes due to chronic stasis of intraluminal contents in the dilated hypo-motile segment. Local mesenteric lymphadenopathy was assumed to be reactive, although the possibility of bowel lymphoma was
raised. The lack of a definitive diagnosis, the localization of the process, and the possibility of malignancy resulted in surgical intervention. Pathology of the resected jejunal segment established the final diagnosis of HGM.

HGM was first described by Schmidt in 1805 [1]. It is classified as either congenital (heterotopic) or acquired (metaplastic). HGM is more commonly found in Meckel’s diverticulae and gastrointestinal duplications [2]. However, it has been reported throughout the entire alimentary tract from the oral cavity to the anus, in the airways, umbilicus, urinary bladder, and even in the scrotum [1–11]. Occasionally, HGM is an incidental pathological finding. However, it is symptomatic in the majority of cases. The clinical presentation varies and depends on the size and location of the HGM. HGM can form an intraluminal mass and cause airway or intestinal obstruction [1,4]. It may serve as the lead point for the development of intussusception [6,12–14]. Intestinal mucosal ulceration with GI bleeding is a known complication of HGM [9,15,16]. Intestinal perforation and fistulization to adjacent structures have also been described [17–19].

In all reported cases of HGM in the small intestine the definitive diagnosis was established by histopathological examination of the surgically removed specimens; and in the majority of cases surgery was performed for acute complications such as GI hemorrhage and intestinal obstruction. It is unusual for HGM to manifest as a chronic illness without overt mechanical obstructive or hemorrhagic signs as was the case in our patient. One case was found in the English literature with a near-similar clinical presentation to our case [20]. In the mentioned report, Kobayashi presented a 14-year-old boy with a recurrent volvulus and chronic abdominal pain; HGM was found in a dilated segment of ileum; although recurrent volvulus is a reasonable explanation for this boy’s chronic abdominal pain, ulcerative process resulted from the HGM could also be a cause [20]. The association of HGM with segmental dilatation of the intestine has been reported [6,16,20–22]. Some reports considered segmental dilatation of the intestine and the presence of HGM to be of one embryological origin [5,6,16,21,22]. Kobayashi postulated that segmental bowel dilatation is a sequela rather than an association of HGM; we concur that segmental bowel dilatation in patients with HGM is likely the result of the chronic irritative and ulcerative process caused by the HGM [20]. In our patient, this segmental jejunal dilatation was not seen on initial contrast studies performed at the age of one year, but became apparent with time.

Non-bleeding peptic ulcers in the small intestine are difficult to suspect and to diagnose. This is due to the rarity of the disorder and to the inability to perform a standard endoscopy on the small bowel, although capsule endoscopy is a viable and safe option [23].

3. Conclusion

HGM has a variable clinical course. Chronic symptoms are rare. Peptic ulcers in the small bowel should be suspected when an extensive workup of chronic abdominal pain, vomiting, or diarrhea does not reveal a definite cause. The absence of fecal blood on repeated testing does not rule out intestinal peptic ulcers. The finding of symptomatic segmental dilatation of the intestine should alert the physician to the possibility of the presence of HGM in the dilated segment causing the symptoms. In such cases a preoperative 99mTc pertechnetate scan or capsule endoscopy can be helpful; however, only pathologic review can establish the definitive diagnosis.

Acknowledgment

Dr. Walid Khalbus, professor of pathology, for his contribution in establishing the final diagnosis, and for providing the histopathology pictures.

Dr. Anna Shawyer, Pediatric Surgeon at McMaster Children’s Hospital, for assistance with editing of the manuscript.

References


