Radiology Case Reports 13 (2018) 839–842

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Side-to-side bowel anastomosis mimicking intussusception in a 2-year-old child with Peutz-Jeghers syndrome☆

Stephanie A. Zacharias[a], *Rajeev Prasad*[a,b], *Faaiza Kazmi*[a,b], *Harpreet Pall*[a,b,*]

[a] *St. Christopher's Hospital for Children, 160 E. Erie Ave, Philadelphia, PA 19134, USA*
[b] *Drexel University College of Medicine, 2900 W Queen Ln, Philadelphia, PA 19129, USA*

ARTICLE INFO

Article history:
Received 9 April 2018
Revised 19 May 2018
Accepted 23 May 2018

Keywords:
Intussusception
Peutz-Jeghers syndrome
Anastomosis
Target sign
Ultrasound
Intestinal polyp

ABSTRACT

Peutz-Jeghers syndrome (PJS) is a rare autosomal dominant inherited condition characterized by hamartomatous gastrointestinal polyps, mucocutaneous pigmentation, and a predisposition for malignancy. Most patients with PJS are diagnosed in the second or third decade of life, and commonly have intussusception as a complication. This report describes an unusual case of a 2-year-old male known to have PJS, who had previously developed a small bowel intussusception caused by a polyp requiring a very short segmental small bowel resection. The patient remained asymptomatic several months after the surgery and then presented with acute abdominal discomfort. On abdominal ultrasound, a target sign measuring 2.7 cm was noted in the left upper quadrant of the abdomen, suggesting a small bowel-small bowel intussusception. There was no evidence of intussusception or bowel obstruction otherwise on diagnostic laparoscopy. It was thought that the previous side-to-side anastomosis had mimicked intussusception on the ultrasound examination. A repeat abdominal ultrasound was performed 1 week after the laparoscopy when the patient was asymptomatic. This again demonstrated a target sign identical in appearance to the previous ultrasound and confirmed that the side-to side anastomosis had in fact mimicked intussusception. It is important that the pediatric gastroenterology, radiology, and surgery communities are aware of this ultrasound finding; it could impact the decision on whether to operate emergently. To our knowledge this is the first report describing this unusual scenario in humans.

Abbreviations: PJS, Peutz-Jeghers syndrome.

☆ Declaration of interest: None.

* Corresponding author.
E-mail addresses: Stephanie.A.Zacharias@gmail.com (S.A. Zacharias), Rajeev.PrasadMD@tenethealth.com (R. Prasad), Faaiza.Kazmi@tenethealth.com (F. Kazmi), hp332@drexel.edu (H. Pall).

https://doi.org/10.1016/j.radcr.2018.05.012

Introduction

Peutz-Jeghers syndrome (PJS) is a rare autosomal dominant inherited condition characterized by hamartomatous gastrointestinal polyps, mucocutaneous pigmentation, and a predisposition for malignancy [1]. Most patients with PJS are diagnosed in the second or third decade of life [2]. However, one-third may be symptomatic by the age of 10 years [3], with an estimated cumulative intussusception risk of 15% [4]. This case report describes a 2-year-old boy known to have PJS who previously developed a small bowel intussusception caused by a polyp requiring a very short segmental small bowel resection [5]. When this toddler was symptomatic several months after surgery, repeat abdominal ultrasound showed a target sign, indicating a small bowel-small bowel intussusception. However, diagnostic laparoscopy showed no evidence of intussusception or bowel obstruction. The target sign was again appreciated on abdominal ultrasound 1 week after surgery when the patient was asymptomatic. To our knowledge, there are no reports that describe this unusual scenario of a stapled side-to-side anastomosis mimicking a target sign on abdominal ultrasound in humans.

Case report

In our practice we care for a 2-year-old boy known to have PJS. This patient was initially diagnosed at 6 months of age based on the pathology of a rectal polyp that was removed after an original concern of rectal prolapse. On subsequent evaluation by esophagogastroduodenoscopy, 2 antral polyps were discovered and removed. At 16 months of age, the patient developed a small bowel intussusception with a 4.2 cm polyp serving as the lead point (Figs. 1A and 2A). Due to the size of the polyp, a very short segmental small bowel resection with a stapled side-to-side anastomosis was required [5]. He remained asymptomatic for 8 months following this operation. At 24 months of age he presented to our outpatient gastroenterology clinic with a 2-day history of abdominal discomfort and constipation in the setting of 1 week of increased fussiness. The recurrent onset of symptoms was concerning given his extensive medical and surgical history. At the time, his vital signs were stable, and physical exam was unremarkable. An abdominal X-ray demonstrated a paucity of bowel gas centrally with a few loops of small bowel appearing mildly dilated with a single air-fluid level, possibly indicating an early intestinal obstruction (Fig. 3). On abdominal ultrasound, a target sign measuring 2.7 cm was noted in the left upper quadrant of the abdomen, suggesting a small bowel-small bowel intussusception (Figs. 1B and 2B).

Given his history of Peutz-Jeghers polyps causing intussusception requiring operative intervention, the patient was urgently taken to the operating room for a diagnostic laparoscopy, reduction of the presumed intussusception, and possible further bowel resection. During laparoscopy, the intestine was examined from the ligament of Treitz to the ileocecal valve with no abnormality noted. Specifically, there was no evidence of intussusception or bowel obstruction. The prior anastomosis, in the left upper quadrant, appeared healthy and patent. Therefore, it was thought that the side-to-side anastomosis had mimicked an intussusception on the ultrasound examination. The patient tolerated the procedure well and was discharged the following day. A repeat abdominal ultrasound was performed about 1 week after the laparoscopy when the patient was asymptomatic (Figs. 1C and 2C). The sonographic images again demonstrated a target sign identical in appearance to the previous ultrasound, confirming that the side-to-side anastomosis had in fact mimicked intussusception.

Discussion

This report describes a rare case of a child previously diagnosed with PJS at an unusually young age who had already experienced complications of this rare condition. By 16 months of age, this patient had underwent a very short segmental

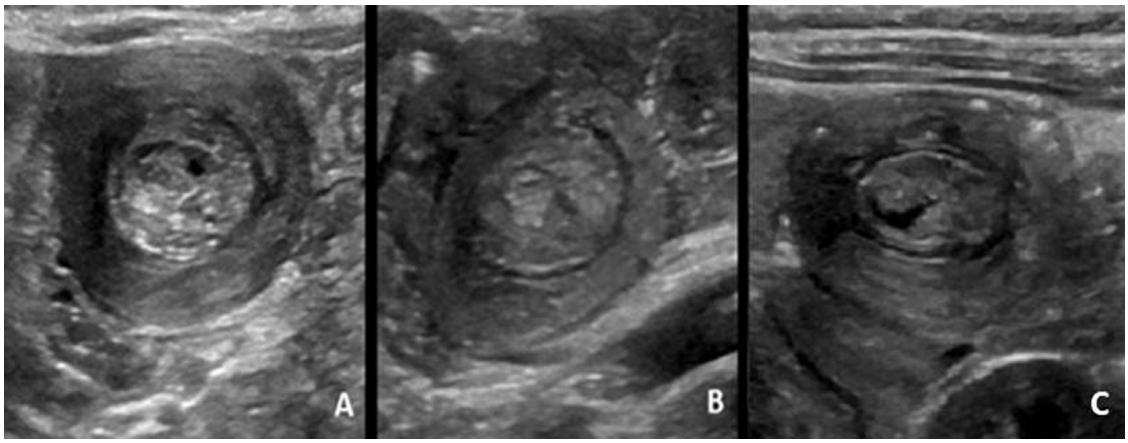

Fig. 1 – Ultrasounds of the left upper quadrant of the abdomen transverse to the axis of the bowel showing a target sign indicating intussusception. (A) Before first surgery. (B) Before second surgery. (C) One week after second surgery when patient was clinically asymptomatic.

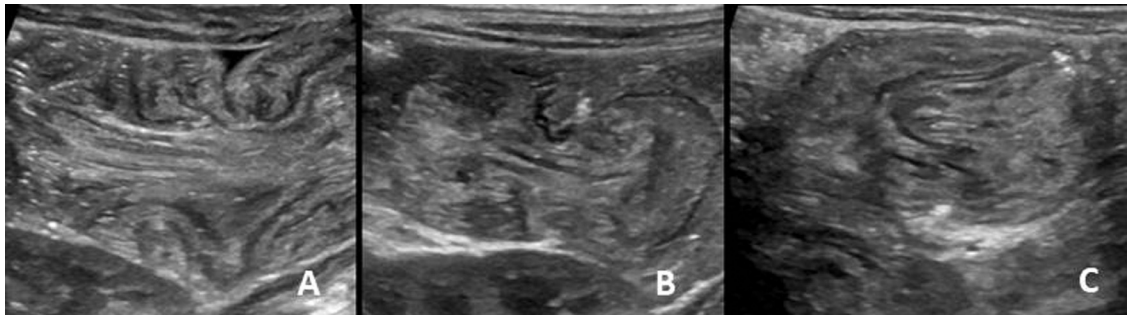

Fig. 2 – Ultrasounds of the left upper quadrant of the abdomen longitudinal to the axis of the bowel indicating intussusception. (A) Before first surgery. (B) Before second surgery. (C) One week after second surgery when patient was clinically asymptomatic.

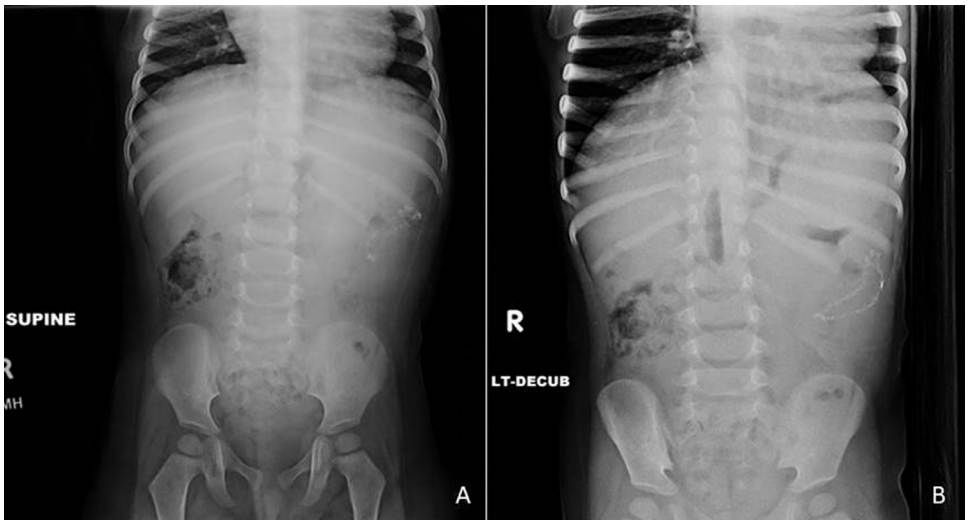

Fig. 3 – Abdominal X-rays before the second surgery. There is a paucity of bowel gas centrally with a few loops of small bowel appearing mildly dilated with a single air-fluid level, possibly indicating an early intestinal obstruction. Note the very small titanium staples in the upper left quadrant of the abdomen. (A) Supine. (B) Left decubitus.

small bowel resection with a stapled side-to-side anastomosis after developing a small bowel intussusception secondary to a polyp that served as a lead point [5]. Due to this previous medical and surgical history, there was a heightened concern for a subsequent intussusception in this patient.

Intussusception occurs when a more proximal portion of intestine (the intussusceptum) telescopes into another more distal portion of intestine (the intussuscipiens). On ultrasound imaging, in transverse view, an intussusception has the appearance of a target due to a central hyperechogenicity consisting of the intussusceptum with its associated mesenteric fat surrounded by a more hypoechoic peripheral rim representing the edematous intussuscipiens. Although ultrasound has a high sensitivity and specificity in diagnosing intussusception, in this case the patient's prior side-to-side anastomosis appears to have mimicked a target sign. It is not entirely clear what anatomic features of the anastomosis resulted in the target sign appearance; however, we postulate that the central echogenic appearance of the "target sign mimic" is due to the very small titanium staples connecting the 2 bowel walls together as part of the stapled side-to-side anastomosis.

In the literature, this "target sign mimic" has been described in dogs in various scenarios [6], but there is no literature on humans. In dogs these mimics typically have a thinner, ill-defined outer wall of the intussuscipiens, which is an important differentiator of intussusception from healthy tissue. However, there were no sonographic features that could help differentiate the mimic in our case from a true intussusception because images within the same study showed variations in appearance.

It is important that the pediatric gastroenterology, radiology, and surgery communities are aware of this ultrasound finding. To our knowledge, this is the first report describing this unusual scenario in humans. Knowing about this radiologic mimic has the potential to impact clinical decision making. If this patient were to present to our hospital again with similar symptoms and imaging studies, he would be admitted for observation rather than being immediately sent to the operating room. In order to better apply this unique mimic

to other patients, further investigational studies would be needed. For instance, a study team could look for target signs in patients with a known previous bowel anastomosis who are undergoing an ultrasound for another reason. If a "target sign mimic" were also found in those patients, then the ultrasound finding in this case would likely be applicable in other cases and could impact a surgeon's decision on whether to operate emergently.

REFERENCES

[1] Jeghers H, McKusick VA, Katz KH. Generalized intestinal polyposis and melanin spots of the oral mucosa, lips and digits; a syndrome of diagnostic significance. N Engl J Med 1949;241(26):1031–6. doi:10.1056/NEJM194912292412601.

[2] Utsunomiya J, Gocho H, Miyanaga T, Hamaguchi E, Kashimure A. Peutz-Jeghers syndrome: its natural course and management. Johns Hopkins Med J 1975;136:71–82.

[3] Beggs AD, Latchford AR, Vasen HF, Moslein G, Alonso A, Aretz S, et al. Peutz-Jeghers syndrome: a systematic review and recommendations for management. Gut 2010;59(7):975–86. doi:10.1136/gut.2009.198499.

[4] van Lier MG, Mathus-Vliegen EM, Wagner A, van Leerdam ME, Kuipers EJ. High cumulative risk of intussusception in patients with Peutz-Jeghers syndrome: time to update surveillance guidelines? Am J Gastroenterol 2011;106(5):940–5. doi:10.1038/ajg.2010.473.

[5] Zacharias SA, Prasad R, Ciullo S, Mallon MG, Marinovich A, Pall H. Small bowel intussusception in a 16-month-old child with Peutz-Jeghers syndrome. *Clin Pediatr* 2017 9922817732148. doi:10.1177/0009922817732148.

[6] Patsikas MN, Papazoglou LG, Papaioannou NG, Dessiris AK. Normal and abnormal ultrasonographic findings that mimic small intestinal intussusception in the dog. J Am Anim Hosp Assoc 2004;40(2):147–51. doi:10.5326/0400147.