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Henoch–Schönlein Purpura Presenting with Intussusception in a 14-Year-Old Female: A Case Report

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Reported here is the case of a 14-year-old female with Henoch–Schönlein purpura (HSP) who presented with a two day history of intermittent abdominal pain and vomiting. Examination demonstrated abdominal guarding. Plain abdominal radiography suggested bowel obstruction, and ultrasound confirmed an ileo ileal intussusception with preserved vascular flow. She underwent an exploratory laparotomy with manual reduction of the ileo ileal intussusception. Her postoperative course and subsequent follow up were unremarkable. This case illustrates how early, accurate diagnosis—achieved through thorough clinical assessment and targeted ultrasonography—is essential for timely intervention. It also highlights the need for prompt surgical intervention to prevent severe complications and the importance of combining clinical vigilance with appropriate diagnostic modalities to optimize patient outcomes.

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Key words: Henoch-Schonlein purpura, IgA vasculitis, bowel obstruction, intussusception, target sign, donut sign

Intussusception in a patient with Henoch-Schönlein purpura (HSP) is a unique and educational case due to its rarity and the specific challenges it presents in diagnosis and management. Intussusception is the most common cause of acute surgical abdomen in children with HSP, occurring in about 5% of cases, yet it remains a rare complication overall. The condition primarily involves the small bowel, such as ileoileal intussusception, which differs from the more common ileocolic intussusception seen in idiopathic cases. This difference requires a high degree of clinical suspicion and the use of appropriate imaging techniques, such as ultrasound, which is highly sensitive and specific for detecting intussusception in HSP patients.[1,3]

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This case is presented to emphasize the critical importance of prompt diagnosis and intervention to prevent potentially fatal outcomes and underscores the need for clinicians to recognize atypical presentations of HSP.

The Case

The patient is a 14 year old female, a known case of Henoch-Schonlein Purpura, diagnosed via skin biopsy in 2018. History started 2 days prior to her admission, when she developed sudden onset of crampy, burning epigastric pain with a severity of 8/10 on the pain scale. This was associated with 1 episode of spontaneous vomiting of previously ingested food, approxiamtely 1cup, non-bilous, non-bloody, non-projectile. No other symptoms were noted. No fever, no changes in bowel habits. No consult was done at this time.

Though intermittent, her epigastric pain persisted. It was non-radiating, crampy, and burning though there was no recurrence of vomiting. At this time, she developed purpuric rashes over her 4 extremities. These were non-blanching and non-pruritic (Figure 1). This prompted consult in another institution wherein she was managed as a case of acid related disease vs acute gastroenteritis. She was sent home stable and was started on oral Cefuroxime, Sodium Alginate+Sodium Bicarbonate+Calcium Carbonate and a Proton Pump Inhibitor (PPI).

The intermittent abdominal pain and rashes persisted. Recurrence of vomiting on the day of admission prompted the patient to seek consult in the emergency room. Abdominal X-ray (Figure 2A) and a complete blood count which were unremarkable. A bacterial predominance was noted on urinalysis. She was advised admission



Figure 1. Multiple purpuric rashes on the bilateral upper and lower extremities.

for observation and for the initiation of intravenous fluids, hydrocortisone, PPI, and Ceftriaxione. While admitted, the patient's epigastric pain and vomiting episodes had resolved. Intravenous hydrocortisone and PPI medications were shifted to oral. She was being prepared for discharge on her third day of admission when her epigastric pain recurred, now radiating to the periumbilical area, crampy, and burning, 8/10 on the pain scale. She had 5 episodes of vomiting. The patient was referred to surgery service for co-management. The surgical service observed that the patient's abdomen was slightly distended with abdominal guarding. Repeat abdominal X-rays revealed signs of obstruction (Figure 2B and 2C). A whole abdominal ultrasound was subsequently conducted, which identified a large echogenic focus measuring approximately 7.5 cm in length in the right lower quadrant, showing the classic "target" or "doughnut" sign and there was also note of preserved vascular flow consistent with intussusception (Figure 3). Findings were interpreted to be compatible with intussusception, likely ileocolic in location.

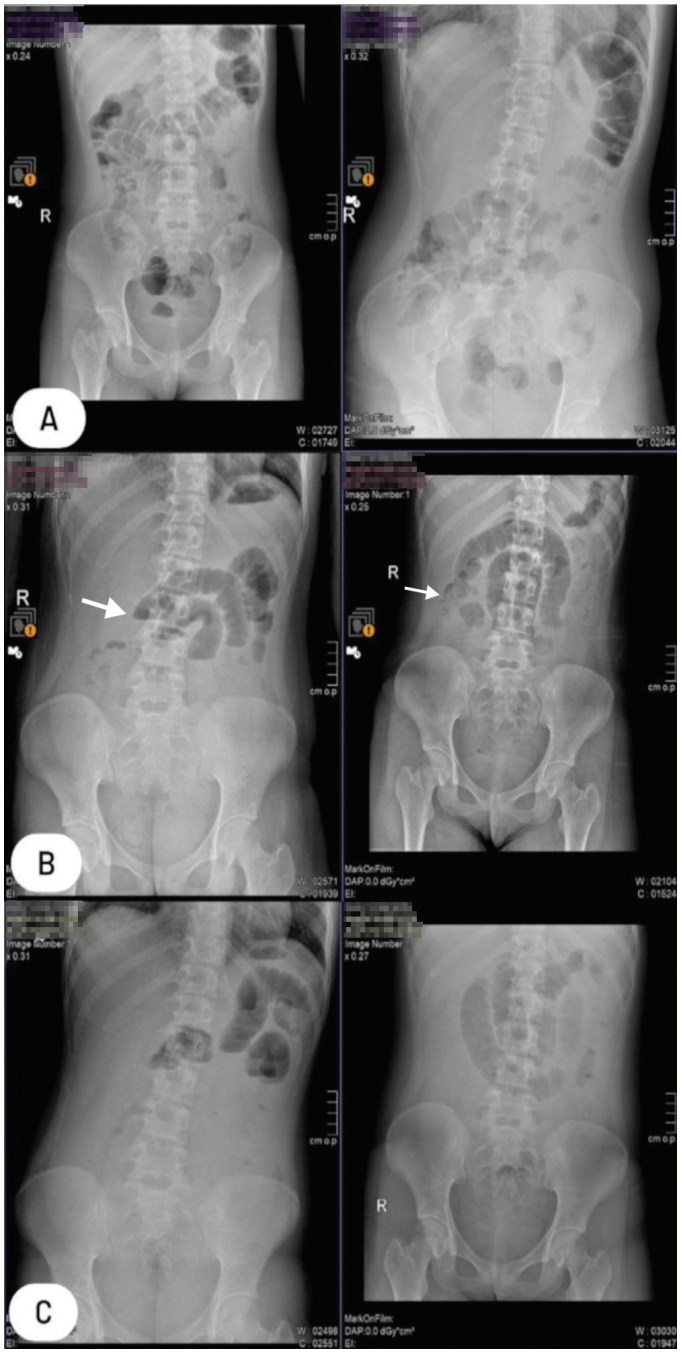


Figure 2. A.) Abdominal X-ray on the first hospital admission with no signs of obstruction. B.) Abdominal X-ray performed on the 3rd hospital day. There were gas-dilated small bowel loops in the central abdomen, with an organized pattern and air-fluid levels (arrows). There was a paucity of distal colonic gas. C.) Abdominal X-ray taken approximately 6 hours later. There were no significant changes in the number and size of gas-dilated small bowel loops in the central abdomen. There is a persistent paucity of distal colonic gas.

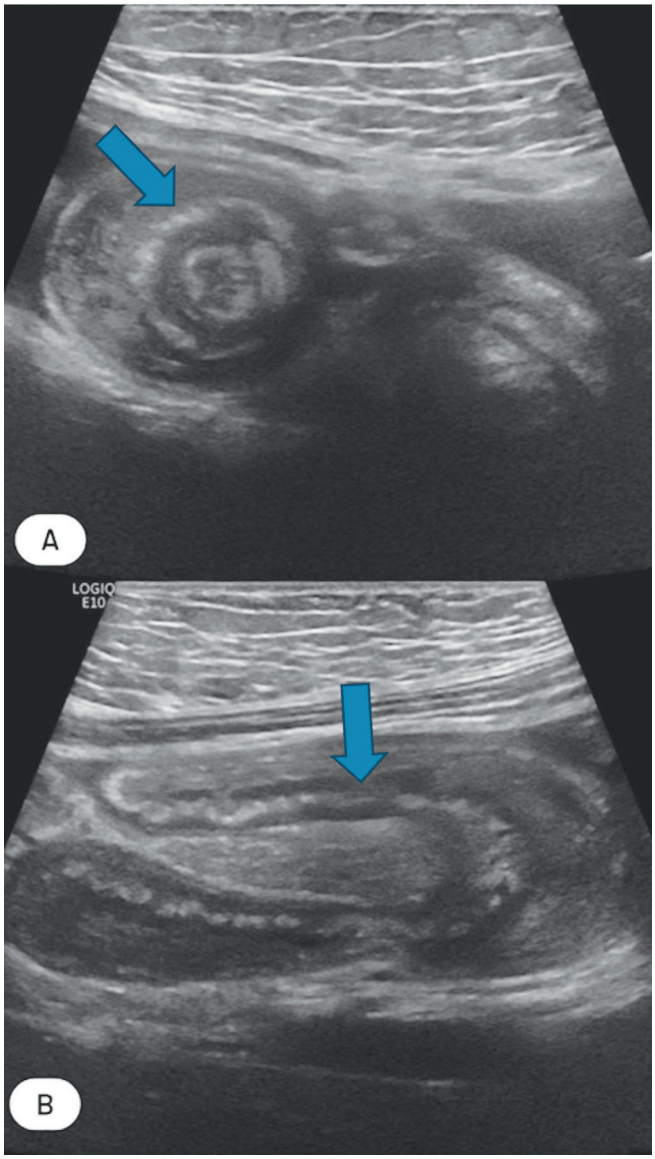


Figure 3. A.) Ultrasound study across the short axis of the abdomen demonstrating the target/doughnut sign (arrow) in intussusception B.) Ultrasound study long axis of the abdomen showing the pseudokidney sign (arrow).

The initial presentation of abdominal pain and vomiting in this patient with HSP raised suspicion for gastrointestinal complications. The guarding noted on physical examination suggested peritoneal irritation, warranting further investigation. The abdominal X-ray indicated obstruction, prompting the use of ultrasound to confirm the presence of intussusception. Other potential diagnoses considered could have included appendicitis, bowel perforation, or other forms of bowel obstruction, but the ultrasound findings were definitive for intussusception. The preserved vascular flow observed on ultrasound was crucial for surgical planning, indicating that the bowel was not yet ischemic.

She was initially managed conservatively. To prevent further gastrointestinal distress, the patient was initially kept *nil per os* (NPO) and was kept hydrated intravenously. However, the patient's symptoms persisted so the decision was made to proceed with an exploratory laparotomy. The procedure was performed by a qualified pediatric surgeon experienced in pediatric abdominal surgeries. The patient was positioned supine. The skin was prepared with a povidone iodine solution. The abdomen was approached through a midline infraumbilical incision carried down to the peritoneum. Upon opening of the peritoneum, approximately 300 cm^3 of ascitic fluid was observed. The proximal small bowel was noted to be dilated. There was note of an ileo-ileal intussusception located 40 cm from the ileocecal valve (Figure 4). The intussusceptum appeared beefy red, while the intussuscipiens was dark red in color with a 4 cm serosal tear on the anti-mesenteric side. After manual reduction, good perfusion was noted in the affected bowel segments (Figure 5). No mesenteric ischemia was observed. The large bowel was unremarkable. The serosal tear was repaired using 3-0 braided polyglactin 910 suture (Vicryl, Ethicon). The abdomen was carefully closed in layers tension-free.

Post-operatively, the patient was gradually reintroduced to oral intake. She was monitored for signs of recurrence or complications. Pain was managed until she recovered and was discharged home.

The patient was discharged on the sixth postoperative day. At the time of discharge, the patient reported tolerable postoperative pain and exhibited no signs of nausea, vomiting, or fever. The patient was instructed to clean the surgical wound with an antiseptic solution and dress it daily. Bowel movements were regular. During the follow-up visit with the surgeon one week post-discharge, the surgical incision was dry and well coaptated. The patient was able to tolerate a full regular diet. No complications or adverse events were reported during the postoperative period.

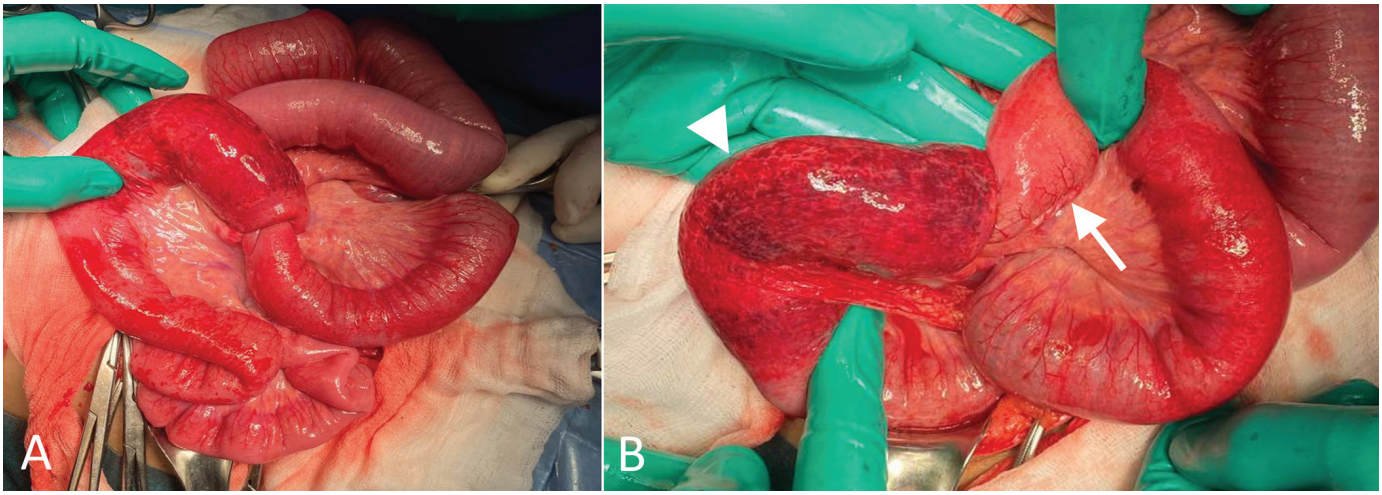


Figure 4: Ileo-ileal intussusception located 40 cm from the ileocecal valve. A.) The bowels proximal to the intussuception were noted to be dilated. B.) The intussusceptum (arrowhead) appeared beefy red, while the intussuscipiens (arrow) was dark red in color.

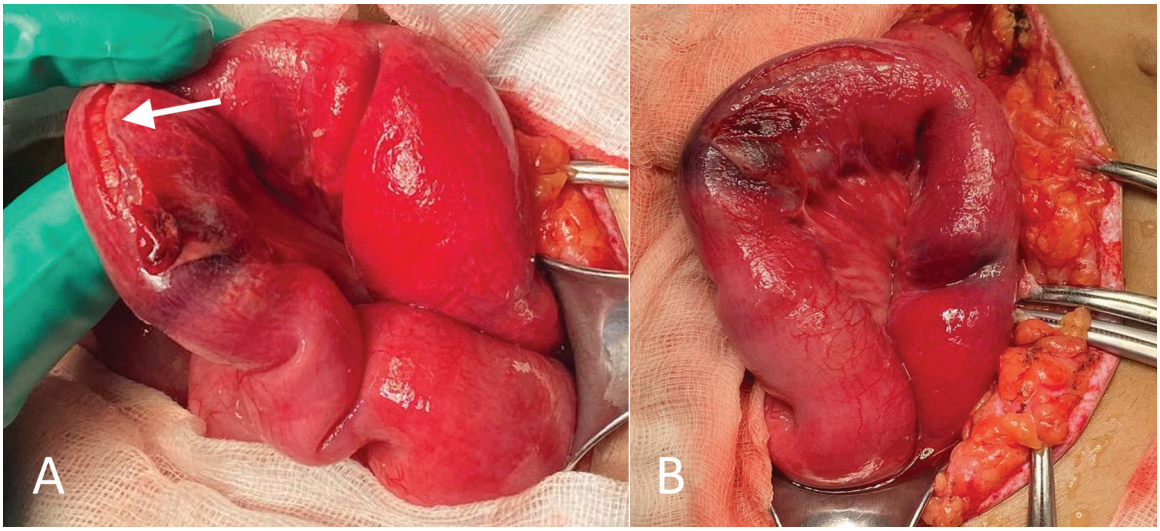


Figure 5: Post manual reduction of the ileo-ileal intussusception. A.) A 4 cm serosal tear (arrow) was noted along the anti-mesenteric side of the intussusceptum. B.) There was good tissue perfusion upon reduction of the intussusceptum with no note of any mesenteric ischemia.

Discussion

Intussusception in HSP is thought to result from IgA-mediated small vessel vasculitis involving the intestinal wall, leading to submucosal edema, hemorrhage, and segmental thickening of the bowel. These changes create a mismatch in caliber and motility between edematous and relatively normal segments of intestine, predisposing the affected bowel to telescope and explaining why small bowel–small bowel intussusception is more common than ileocolic forms in HSP.[1-4] Mesenteric vasculitis may further impair venous return, aggravating congestion and accelerating progression to ischemia once intussusception occurs if not promptly reduced. Abdominal vasculitic involvement in HSP can mimic more benign presentations (colicky pain, vomiting, or gastrointestinal bleeding)

and may be mistaken for nonsurgical causes, delaying recognition of mechanical obstruction.

Ultrasound is appropriately the first line imaging modality in pediatric HSP with abdominal symptoms, given its high sensitivity and specificity for intussusception and absence of ionizing radiation, and it reliably demonstrates the classic "target" and "pseudokidney" signs. In the present case, it not only confirmed ileo-ileal intussusception but also documented preserved vascularity, which was important for operative planning. However, ultrasound can be limited by operator dependence, overlying gas, and difficulty assessing more proximal or multiple small bowel intussusceptions. In selected cases, especially in older children or when ultrasound is equivocal, contrast enhanced computed tomography (CT) may be considered to provide a more global assessment of bowel wall enhancement, length of the intussusception, and signs of ischemia, pneumatosis, or perforation, helping to identify cases requiring urgent surgery while balancing radiation exposure.

The role of non-operative reduction in HSP associated intussusception is more limited than in idiopathic ileocolic disease. Pneumatic or hydrostatic enemas are highly effective for ileocolic intussusception but have lower success rates in small bowel–small bowel involvement and may risk missing necrosis in edematous, vasculitic bowel. For this reason, many authors advocate early surgical involvement and a lower threshold for operative management when HSP patients develop persistent pain, signs of obstruction, peritoneal irritation, or imaging evidence of small bowel intussusception.[1–3,5,11] Laparoscopy can be a useful alternative to immediate open surgery in hemodynamically stable patients. It allows diagnostic inspection and, in selected cases, gentle laparoscopic reduction, with potential benefits including less pain and shorter hospitalization. However, conversion to open laparotomy should be prompt if there is bowel distension, poor visualization, or suspected ischemia. In our case, the recurrence and progression of symptoms, radiographic evidence of small-bowel obstruction, and ultrasound-confirmed ileo-ileal intussusception supported proceeding directly to exploratory laparotomy. Timely manual reduction preserved bowel viability and avoided resection, consistent with recommendations for early operative intervention in complicated HSP-related intussusception.[1–3,5,7,11] This case report illustrates the critical importance of maintaining a high index of suspicion for intussusception in patients with Henoch-Schönlein purpura (HSP) who present with abdominal symptoms. Intussusception, though a rare complication of HSP, can lead to significant morbidity if not promptly diagnosed and treated. A thorough physical examination and continuous monitoring were essential for detecting early signs of abdominal guarding and obstruction, enabling timely ultrasound use for diagnosis.

The literature emphasizes that prompt diagnosis and management are crucial to prevent severe outcomes such as bowel obstruction and necrosis. Ultrasound remains a highly effective diagnostic tool due to its sensitivity and specificity, particularly in pediatric populations with conditions like HSP.[6,8] Surgical intervention, as demonstrated in this case, is often necessary when conservative measures are insufficient, aligning with current clinical guidelines for managing intussusception.[7,9-10]

Conclusion

Early detection and intervention are key in successful management of complex pediatric conditions such as this unusual case of intussusception. It underscores the necessity of clinical vigilance combined with the appropriate diagnostic and therapeutic strategies to achieve favorable outcomes.

Informed Consent

Verbal and written informed consent was acquired from the patient for the use of his information while maintaining anonymity and patient privacy.

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