Intussusception: It's Not Always Ileo-Colic

A 5-year-old boy presented with intermittent episodes of central abdominal pain and vomiting for 3 days. Despite this, he was well hydrated with a soft non-tender abdomen on clinical examination. There was past history of intussusception 8 months prior, which was successfully reduced with fluoroscopic-guided air enema reduction. At presentation, an abdominal ultrasound was performed to investigate the abdominal pain (Fig. 1).

What is the diagnosis?

A) Ileo-colic intussusception
B) Ileo-ileo-colic intussusception
C) Ileo-ileal intussusception
D) Jejuno-jejunal intussusception
E) Colo-colic intussusception

Findings and Diagnosis

The ultrasound images (Fig. 1) show a mass-like lesion demonstrating a "target sign" in the middle left abdomen. Normal vascularity is preserved within the walls of the intussusceptum and intussuscipiens. There is a polypoid structure with a vascular stalk seen within the intussusceptum, consistent with a polyp. The caecum and appendix are noted to be in normal position, in the right iliac fossa. The overall features are compatible with colo-colic intussusception with a polyp acting as the lead point.

Subsequently, the patient underwent successful reduction of the colo-colic intussusception using air enema procedure. Further evaluation with colonoscopy and endoscopic resection revealed a 2.8 cm juvenile polyp at the junction of the descending and sigmoid colon.

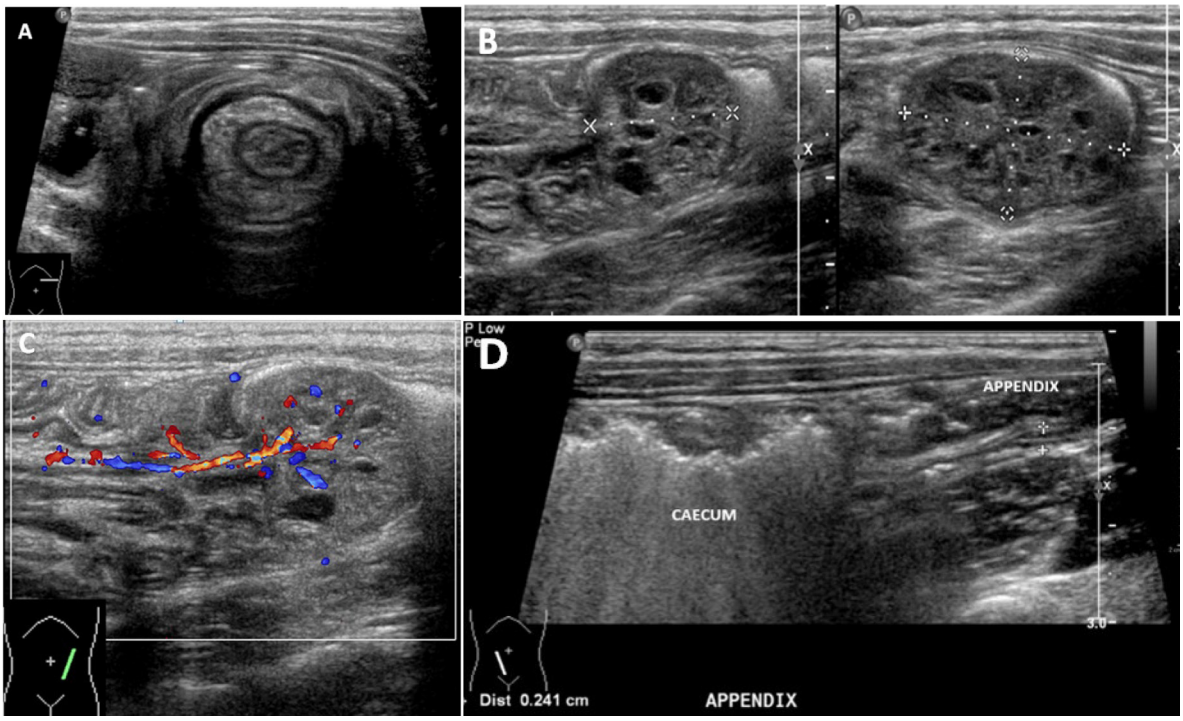

Fig. 1. A) A mass-like bowel lesion in the left upper quadrant of the abdomen with a "target sign" is seen. B & C) A polypoid mass with a vascular pedicle is seen within the intussusceptum. D) Caecum and appendix are detected at normal position in the right iliac fossa.

Answer: E

Discussion

Intussusception is the telescoping of a segment of bowel (the intussusceptum) into another segment of the bowel (the intussuscipiens).[1] It is the most common cause of bowel obstruction in the paediatric age group, occurring most commonly between the ages of 6 months and 2 years.[2] In our case, the child was above the typical age group for intussusception and has experienced recurrent episodes of intussusception. This raises the suspicion of a pathological lead point as a cause of the intussusception.

The classical triad of colicky abdominal pain, vomiting and passage of "red currant jelly" stool is present in less than 50% of cases.[3] A 3-year review by Lai et al demonstrated that 56.3% of patients presented with a palpable abdominal mass.[4] Early diagnosis is crucial for adequate management and prevention of complications. If left undiagnosed and untreated, bowel infarction or perforation may occur.

Plain abdominal radiographs have a limited role in diagnosis of the intussusception due to its low sensitivity (48%) and specificity (21%).[5] However, they are helpful in the detection of complications such as small bowel obstruction and perforation.

Ultrasound is the diagnostic modality of choice, due to its high sensitivity (97.9%) and specificity (97.8%),[5] with a high negative predictive value of 100%.[3] Abdominal sonography may demonstrate a soft tissue mass comprising alternating hyperechoic and hypoechoic rings, known as the "target" or "doughnut" or "bull's eye" sign.[3] "Crescent-in-doughnut" sign is the presence of a hyperechoic crescent-like area at the centre of the alternate hyperechoic and hypoechoic rings, due to the invagination of the mesenteric fat.[3] In addition, mesenteric lymph nodes can be detected within the crescentic mesenteric fat. Occasionally, a pathological lead point can be identified on ultrasound. Navarro et al showed in a study that ultrasound can detect 64% of the pathological lead points.[6] Preservation of vascularity within the intussusception can be observed on colour Doppler. Small amount of free intraperitoneal fluid is frequently seen on ultrasound.

Computed tomography and magnetic resonance imaging are not routinely used to diagnose intussusception in children although both are highly accurate in the diagnosis of the condition. These modalities are reserved for evaluation of selected patients with an atypical sonographic appearance, suggesting a pathological lead point such as lymphoma or a bowel mass.

Treatment options include image-guided enema reduction and surgical reduction. Contraindications for non-surgical management are dehydration, haemodynamic instability, peritonitis and perforation on abdominal X-ray.[7] The 2 most common techniques for image-guided enema reduction are fluoroscopy-guided pneumatic reduction and ultrasound-guided hydrostatic reduction. Overall, a success rate of 80% to 95% can be achieved by image-guided enema reduction.[7] Complications of image-guided reduction include perforation, with an overall rate of less than 1%.[7] Recurrence of the intussusception can occur in approximately 10% of cases, in which 50% will occur within 48 hours after initial reduction.[7,8] If image-guided reduction is not successful after 3 attempts, surgical reduction may be required.

There are different types of intussusception, depending on the region of the affected bowel; these include ileo-colic, ileo-ileo-colic, colo-colic and small bowel (jejuno-jejunal and ileo-ileal) intussusception.[3] The most common type is the ileo-colic variant, without a pathological lead point seen in a majority of the cases. According to Blake Lock et al, intussusception in children with a pathological lead point as a cause ranges from 1.5% to 12%.[9]

In the literature, the frequency of small bowel intussusception (ileo-ileal) varies between 1.6% to 25% of all cases of intussusception. Most of these cases are transient[10] and are treated conservatively.

Colo-colic intussusception is an unusual variant of a common paediatric condition, with only a few cases reported in the current literature. Review of 5 articles—published between 1978 to 2010 by Takahashi et al—showed that the mean age at diagnosis was 9.5 years (range, 7-11 years), with an increased prevalence in females.[11]

Most paediatric colo-colic intussusception cases occur with a lead point such as polyp(s)[12] or neoplasm (lymphoma).[3] Other rare causes reported in the literature have occurred in the clinical setting of Henoch-Schonlein purpura,[8] inflammatory bowel disease, adenocarcinoma, ganglioneuroma and hereditary angioneurotic oedema.

Image-guided enema reduction may be difficult in the presence of a pathological lead point, with a lower success rate and higher chance of recurrence. After successful reduction, further investigation with colonoscopy is recommended to evaluate the nature of the pathological lead point. Thus, it is important to accurately diagnose the colo-colic intussusception initially.

Ultrasound can help differentiate between ileo-colic and colo-colic intussusception. In ileo-colic intussusception, the normal caecal pole is not identified in the right iliac fossa due to invagination of the ileum through the ileocaecal valve into the caecum/colon. With careful scanning, the appendix may also be seen within the intussusceptum (Fig. 2). In contrast, in colo-colic intussusception (as in our case), the caecal pole and appendix remain at the normal position in the right iliac fossa (Fig. 1). In addition, the presence of a pathological lead point such as a polyp within the intussusceptum raises suspicion of a colo-colic intussusception.

In our case, the ultrasound features demonstrate the target sign, consistent with an intussusception. A polypoidal structure, a polyp with a vascular pedicle was seen within the intussusceptum. The appendix and caecum were noted in the right iliac fossa, thus excluding an ileo-colic intussusception.

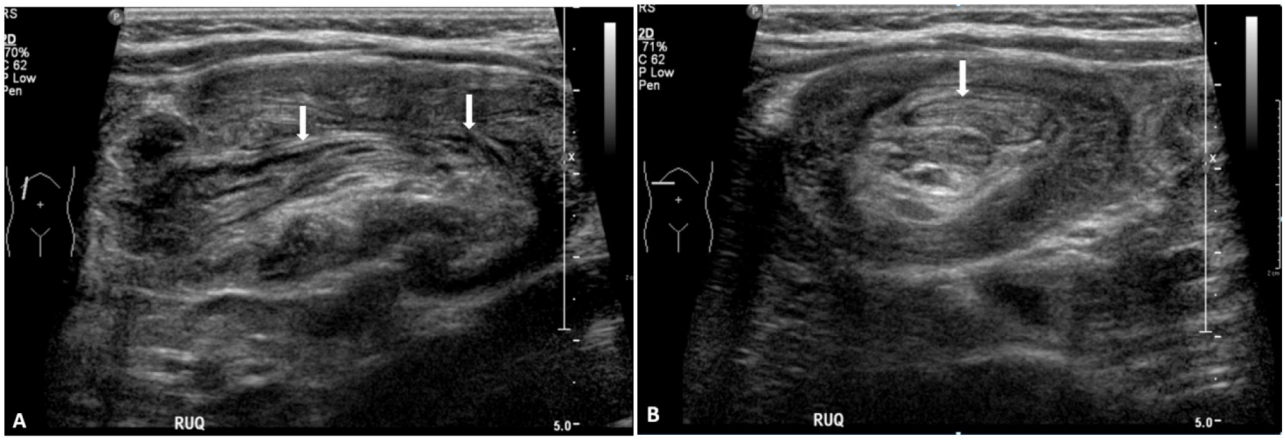

Fig. 2. Typical ileo-colic intussusception in the right upper quadrant of the abdomen. A) Longitudinal view shows a linear tubular structure (arrows) within the intussusceptum in keeping with the appendix. B) On transverse view, tip of the appendix (arrow) can be seen.

The diagnosis was that of colo-colic intussusception, with a polyp as a pathological lead point. It was successfully reduced with fluoroscopic-guided air enema reduction and no immediate complications occurred. The presence of a polyp at the junction of the descending and sigmoid colon was confirmed on colonoscopy performed at a later date (Fig. 3) and this was confirmed to be a juvenile polyp on histology.

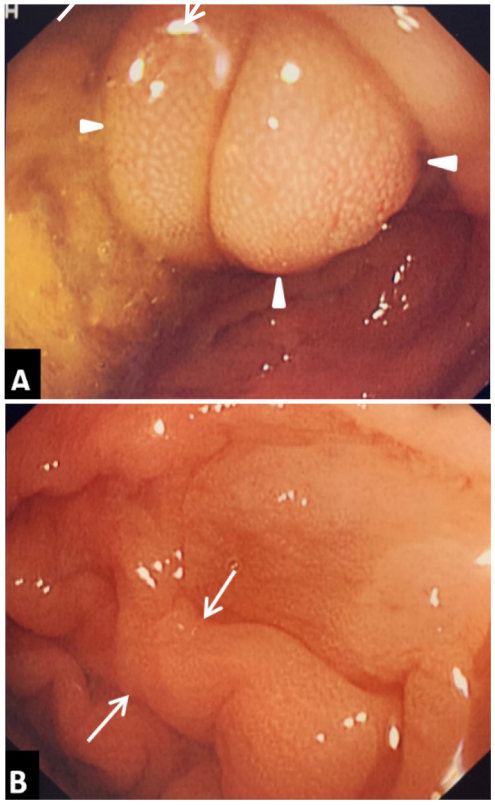

Fig. 3. Colonoscopy images show a polyp at the junction of the descending and sigmoid colon (arrowheads in A) with a stalk (arrows in B).

Conclusion

Intussusception is the most common cause of paediatric bowel obstruction. There are different types of intussusceptions. Most cases of ileo-colic intussusception are idiopathic in aetiology. The colo-colic intussusception is invariably associated with a pathological lead point, which may result in difficulty in its reduction as well as a higher chance of complications and recurrence. We have demonstrated that in older children (above the typical age group), with recurrent intussusception, a pathological lead point is usually the cause of the intussusception. Ultrasound imaging is crucial not only in the initial diagnosis of intussusception, but also in the accurate differentiation between the different types of intussusception as well as identification of a pathological lead point. In this way, the patient can be adequately managed without significant delay.

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