CASE REPORT

Idiopathic Localised Dilatation of the Ileum: Computed Tomography Enteroclysis

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ABSTRACT
Idiopathic localised dilatation of the ileum is a rare condition in which one or more segments of the ileum are dilated with abrupt transition to normal segments proximally and distally. To the best of our knowledge, this is the first case report to describe the findings and highlight the potential advantages of computed tomography enteroclysis to assess this condition.

Key Words: Diverticulum; Intestine, small; Tomography, X-ray computed

中文摘要

特發性局部迴腸擴張：電腦斷層掃描灌腸造影

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特發性局部迴腸擴張是一種罕見疾病，指迴腸的一個或多個分段擴張而其近端和遠端與正常腸段吻合。以我們所知，這是該病首例報告，文中描述該病的電腦斷層掃描灌腸造影特徵及強調灌腸造影評估該病徵的潛在優勢。

INTRODUCTION

Idiopathic localised dilatation of the ileum (ILDI) is also termed ileal dysgenesis, segmental mega-ileum, and giant Meckel's diverticulum. In this condition, there is one or more short segmental dilatation of the ileum with abrupt transition to a normal bowel lumen proximally and distally. Less than 50 such cases have been reported in the world literature so far. The findings in different imaging modalities were reported but those of computed tomography (CT) enteroclysis have not been previously described. We describe CT enteroclysis appearances of two adult patients with ILDI causing recurrent gastrointestinal bleeding. This case report also highlights the potential advantages of CT enteroclysis (imaging after contrast infusion into the small intestine) in diagnosing this rare disease entity.

CASE REPORTS
Case 1
A 28-year-old man with good past health, presented with melena and mucus in the stool for 1 week, in January 2008. He had no abdominal pain,

weight loss, or change of appetite. He was not on any long-term medication. Physical examination revealed pallor and tarry stool per-rectum. Upon admission, he had anaemia (haemoglobin, 90 g/l). Upper gastrointestinal endoscopy, colonoscopy, and a Meckel's scan were all normal. CT enteroclysis was performed by manual injection of 2000 ml of 0.5% methylcellulose via an endoscopically placed nasojejunal tube. No bowel preparation or conscious sedation was given before the enteroclysis, but 20 mg hyoscine (Buscopan; Boehringer Ingelheim GmbH, Ingelheim, Germany) was given as an anti-peristaltic before the scan. Non-contrast and contrast-enhanced (60 s) CT examinations of the abdomen and pelvis were performed in supine position, and showed a well-demarcated segment (8 cm in length) of axially dilated ileum (4 cm in diameter) in the right lower abdomen

(Figure 1). This bowel segment yielded normal mucosal enhancement and wall thickness, but there was paucity of valvulae conniventes as compared with the adjacent normal ileum. No bowel wall thickening or mass lesion was detected. There was no stricture or obstructive lesion in the rest of the bowel. There was prompt methycellulose transit through this segment of bowel into the colon. Subsequent retrograde (anally inserted) double balloon enteroscopy of the small bowel revealed erythematous mucosa and a 5-mm ulcer 80 cm from the ileocaecal valve, which corresponded to the site of dilated small bowel demonstrated in the CT. Biopsy of the ulcer and erythematous mucosa showed mild inflammation with regenerative changes. The bleeding subsequently resolved spontaneously, and the haemoglobin level returned to normal (150 g/l) during that admission. Two years later, the patient developed
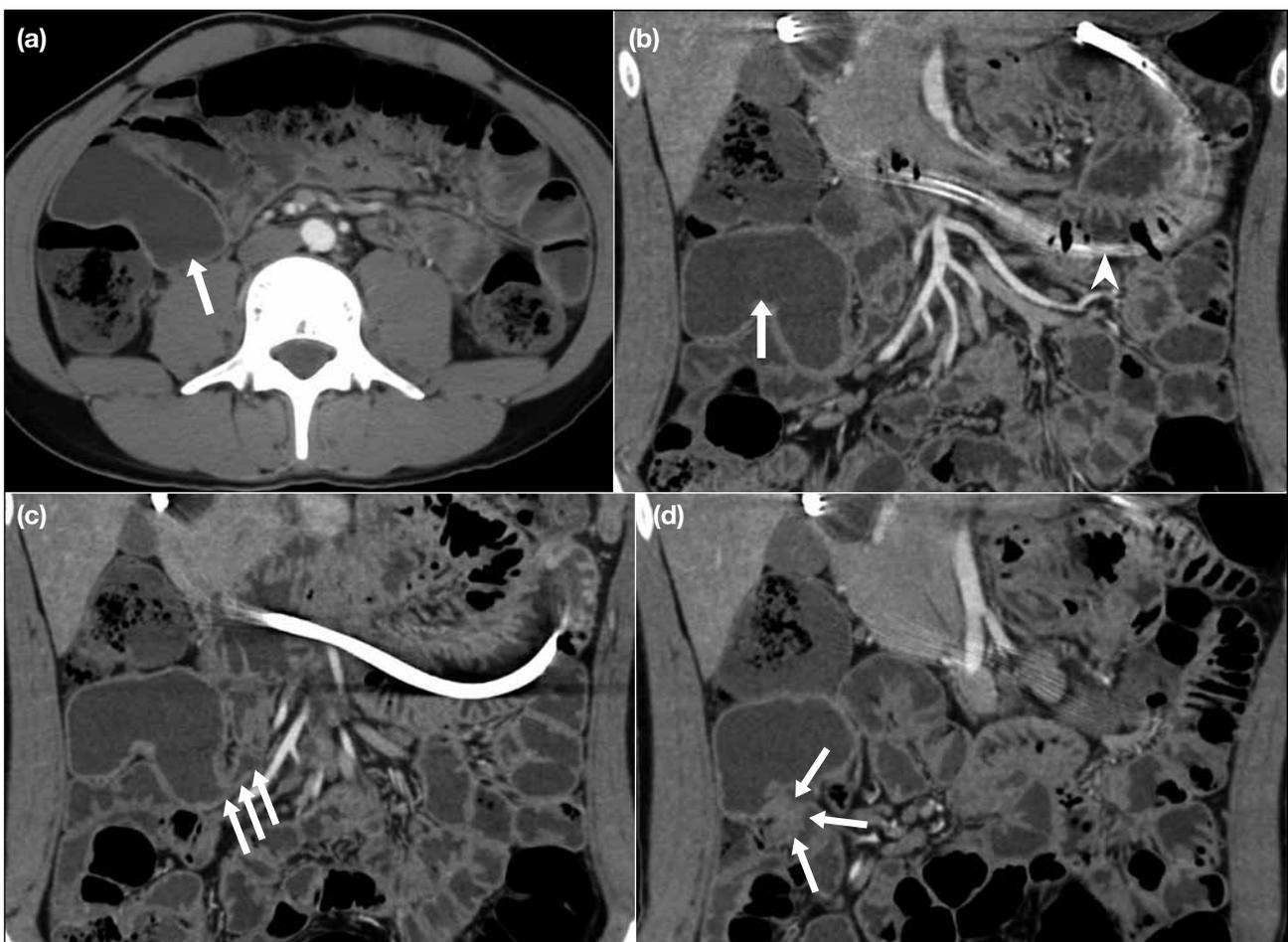
Figure 1. (a) Axial contrast computed tomography (CT) enteroclysis shows a well-demarcated segment (8 cm long) of axially dilated ileum (4 cm in diameter; arrow) in the right lower abdomen. This segment of bowel had normal mucosal enhancement and wall thickness but paucity of valvulae conniventes as compared with the adjacent normal ileum. (b) Coronal contrast CT enteroclysis of this dilated segment of small bowel (arrow). The jejunal catheter for the injection of methylcellulose is revealed (arrowhead). (c) Oblique view of the contrast CT shows abrupt transition to normal ileum proximally (arrows). (d) Oblique view of the contrast CT also shows the abrupt transition to normal collapsed ileum distally (arrows).

another episode of per-rectal bleeding. The bleeding source was not localised by endoscopy or conventional mesenteric angiography. Again the bleeding resolved spontaneously. Follow-up CT enteroclysis 5 years after the first presentation showed no interval change in the appearance of the relevant segment of axially dilated ileum, apart from its slightly different position within the abdomen (Figure 2). Overall, all these features suggested ILDI. The patient refused surgery.

Case 2

A 58-year-old man presented with occult gastrointestinal bleeding, namely iron-deficiency anaemia and positive faecal occult blood test, in October 2011. Upper and lower gastrointestinal endoscopy revealed no abnormality. Subsequently, CT enteroclysis was performed and showed a focal segment of axially dilated distal ileum (7 cm in length and 4 cm at its maximal transverse diameter) [Figures 3a and b]. This dilated ileum showed abrupt transition to normal calibre at the proximal and distal ends (Figures 3a and b). No definite stricture or obstructive lesion was identified. At the mid portion of this bowel segment, there was a focal blind-ending outpouching, suggestive of a diverticulum (Figures 3c and 3d). Overall, the imaging features were suggestive of ILDI with a small Meckel's diverticulum within the dilated ileal segment. The patient refused surgery.

DISCUSSION

The first case of ILDI was reported in 1962,[1] but its aetiology remains unclear.[2] Only a small number
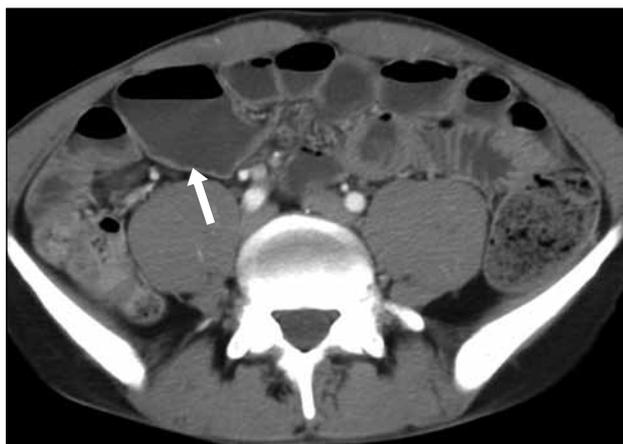
Figure 2. Follow-up contrast axial computed tomography 3 years after initial imaging. There is no significant interval change in this dilated segment of ileum (arrow) although it is in a slightly different position within the abdomen when compared with the previous scan. Overall features are suggestive of idiopathic localised dilatation of the ileum.

of patients manifest in adolescence or adulthood (as in our case).[3] Most occur in children (<10 years old) and there are reported associations with mid-gut congenital anomalies, including exomphalos, malrotation, and Meckel's diverticulum.[2-5] These anomalies suggest the possible congenital nature of ILDI. The proposed aetiologies of ILDI relate to functional or mechanical causes. According to one theory, neuromuscular dysfunction of ileum serves as a functional obstruction resulting in localised dilatation.[2] In another theory, transient gastrointestinal obstruction caused by abdominal wall compression during periods of physiological mechanical uterine herniation or other forms of extrinsic uterine compression has been suggested.[2]

About half of the patients with ILDI present with abdominal pain.[2] Gastrointestinal bleeding is another common complaint. Functional obstructive symptoms due to atonic ileal segment are occasionally noted. In the paediatric group, patients may present with failure to thrive. As in our second case, more than 80% of adults present with occult gastrointestinal bleeding and iron-deficiency anaemia. As in our first case, occasionally, there is massive overt gastrointestinal bleeding. The gastrointestinal bleeding is thought to be related to ulceration caused by chronic stasis of intestinal content or ectopic gastric mucosa. Perforation complicated with peritonitis is a rare presentation.[2]

Pathologically, the involved ileum is thin but otherwise the bowel wall, its nerve plexuses, and ganglion cells are normal.[5] Mucosal ulceration is frequent. Occasionally, heterotopic gastric mucosa is present. In the reported cases, the length of the involved ileal segment ranged from 6 to 21 cm. Tubular, bilobulated, and multilobulated configurations can be found. There may be decreased or absent valvulae conniventes in the involved segment.

The dilated small bowel loop can be detected by plain radiography, even without any evidence of proximal or distal mechanical obstruction.[3] In most instances, small bowel barium studies can illustrate the sharply demarcated segment of axially dilated ileum and paucity of valvulae conniventes.[4] Ulceration may be detected in a minority of cases.[4] Ultrasonography of the involved segment shows up as a fluid-filled cyst-like lesions with or without internal echoes. The real-time detection of peristalsis is helpful in differentiating ILDI from other non-bowel cystic masses.
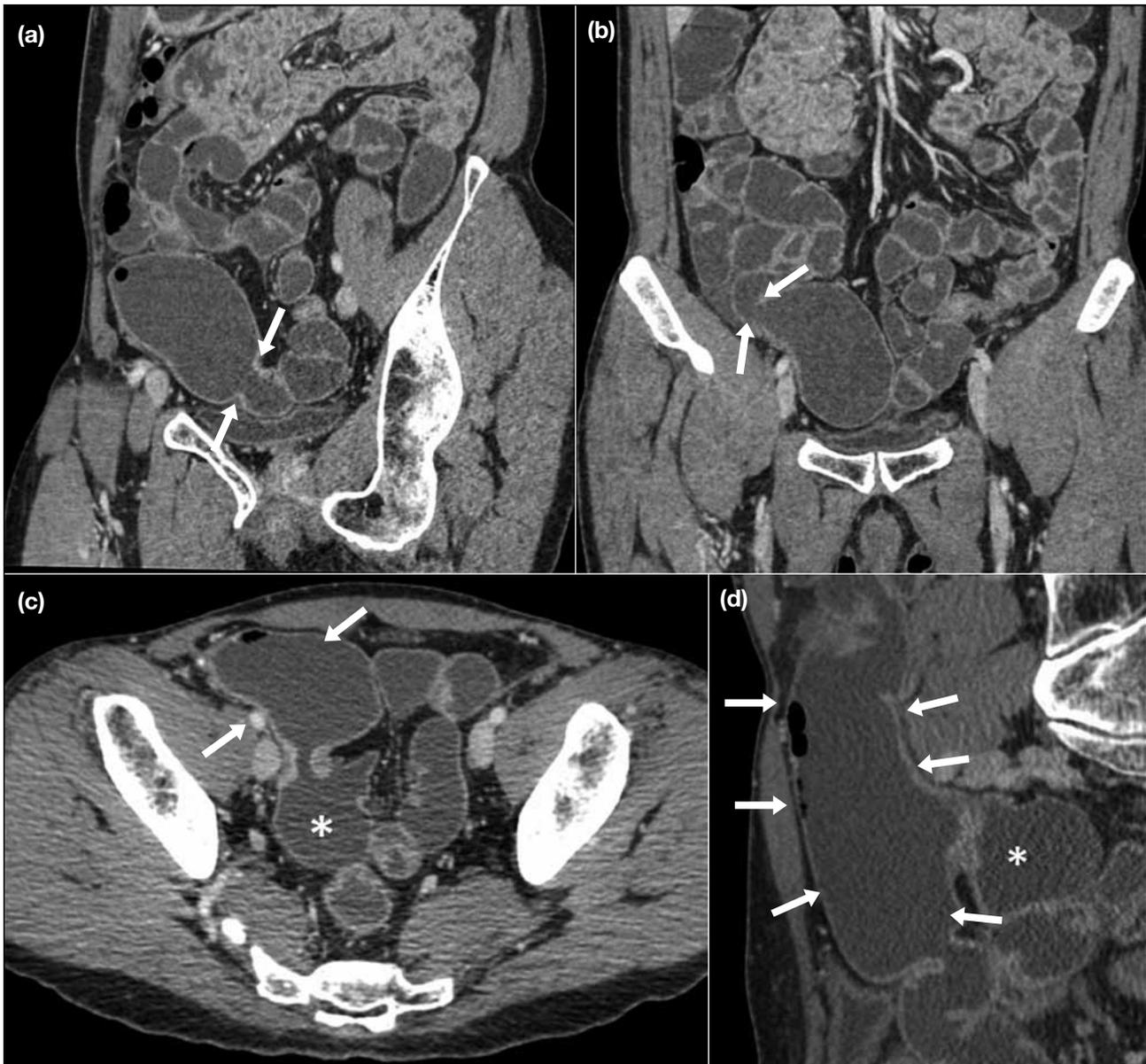
Figure 3. Oblique coronal reformatted images reveal a well-demarcated segment of focal axial dilatation of distal ileum with abrupt transition of calibre both proximally (arrows in a) and distally (arrows in b). In (c) and (d), axial and curved reformatted images show a diverticulum (asterisks) arising eccentrically from the mid portion of axially dilated ileum (arrows). Note the wall thickening with mild mucosal hyper enhancement at the neck of the diverticulum suggests inflammation. The findings are also compatible with localised dilatation of ileum associating with a Meckel's diverticulum.

Two previous reports described the CT findings of ILDI, both detailed segments with localised cystic or tubular bowel dilatation with a thin wall and communication with adjacent bowel loops at two abrupt narrow zones of transition, proximally and distally.[2,3] They also referred to loss of normal valvulae conniventes in the dilated segments. The layering pattern of the bowel wall and mucosal enhancement of the involved segment were similar to the adjacent bowel loops, and the involved segment was axially oriented to the proximal and distal

normal bowel. All of the above findings were identified in our cases. Moreover, CT is superior for excluding neoplastic or inflammatory lesions causing intrinsic or extrinsic narrowing of the bowel.

One previously reported case entailed ILDI missed at laparotomy because the bowel was not distended at time of the operation.[4] Small bowel barium enteroclysis may increase the diagnostic yield of this disease, owing to better bowel distension[4] and is recommended in

patients presenting with occult gastrointestinal bleeding. Compared with small bowel barium examinations with enteroclysis, CT enteroclysis with optimal bowel distension offers clearer visualisation of the configuration as well as the wall of dilated bowel segment. Such imaging is very helpful in making the diagnosis of ILDI. The disadvantage of all plain fluoroscopic small bowel examinations is the inability to provide any extraluminal information.

The differential diagnosis of ILDI includes aneurysmal dilatation of small bowel lymphoma, cavitating neoplasm of the small bowel, Meckel's or other small bowel diverticula, sacculation from scleroderma, scarring in Crohn's disease, postoperative deformity, and dilatation proximal to an obstructing lesion. The irregular thickened bowel wall in lymphoma or a cavitating neoplasm can be easily differentiated from ILDI. Meckel's or other small bowel diverticula should be located along the border of the ileum (i.e. eccentrically rather than axially in relation to adjacent normal bowel). The dilatation in scleroderma is also asymmetrical and non-axial, and the hidebound appearance of the dilated bowel is in marked contrast to the decrease to absence of folds in ILDI. The dilatation due to mechanical obstruction does not show sharp proximal transition (as in ILDI). In Crohn's disease, usually there is bowel wall thickening due to inflammation and other associated findings such as fibrofatty proliferation. Sometimes, postoperative deformity can mimic ILDI, but usually the bowel is more irregularly dilated and there is history of previous bowel surgery.

Angiography may be useful in patients with active bleeding greater than 0.5 ml per minute and can identify highly vascular non-rebleeding lesions such as angiodysplasia and neoplasms.[6] Radionuclide scanning is a non-invasive screening technique and can be used for locating site of acute bleeding, especially from lower gastrointestinal tract if the volume of bleeding is greater than 0.1 to 0.4 ml per minute.[6] Angiography is an invasive procedure and not performed in one of our patients, as both of them were haemodynamically stable. In addition, the bleeding was not massive and resolved spontaneously. Radionuclide scanning may be helpful to localise the site of bleeding, but the information about the cause is usually not obvious. This was why CT enteroclysis was performed instead.

A recent study showed that CT enteroclysis gave a better distension of the jejunum than CT enterography while the ileal distension was similar in both techniques.[7] We postulated that both CT enteroclysis and CT enterography will give similar accuracy in establishing the diagnosis of ILDI. Capsular endoscopy can only reveal the lumen of the small bowel, and pick up associated mucosal inflammation or ulcers, but not dilatation of the small bowel. Capsular endoscopy may not be helpful in establishing the diagnosis of ILDI.

Surgery was not performed in both of our cases as the patients declined operation. Thus, absence of pathological correlation is the main limitation of this case report. However, the CT enteroclysis findings were typical enough for the diagnosis of ILDI to be confidently made. No interval change of the dilatations was evident in the 3-year follow-up CT enteroclysis in case 1, which also supports the diagnosis.

CONCLUSION

Radiologists should be aware of ILDL, which is a rare cause of gastrointestinal bleeding with characteristic imaging features, whilst CT enteroclysis with optimal bowel distension may further increase diagnostic accuracy.

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