Laparoscopic management of Peutz-Jeghers syndrome (PJS) presenting with chronic non-ischaemic jejuno-jejunal intussusception in an adolescent girl

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DESCRIPTION

A 13-year-old girl presented with intermittent upper abdominal pain and vomiting over the duration of 9 months. The physical examination showed pigmentation on the perioral region with upper abdominal fullness and tenderness over left flank. She had iron deficiency anaemia. An abdominal ultrasound scan (USS) showed evidence of non-ischaemic chronic jejuno-jejunal intussusception (figure 1C–E). An upper gastrointestinal contrast study showed dilated duodenal loops and irregularity of bowel in the mid abdomen (figure 1A,B, arrows). During laparoscopy, a large polyp forming the lead point of the intussusceptum in the proximal jejunum was found. Reduction of intussusception, segmental resection of the jejunum containing polyps, resection of the remaining polyps after prolapsing the proximal and distal limbs of the proximal jejunum and primary anastomosis were

performed extracorporeally. The histopathological examination showed hamartoma, confirming the diagnosis of Peutz-Jeghers syndrome (PJS). Her postoperative course was uneventful. She had one colonic and one rectal polyp resected during annual surveillance endoscopies (oesophago-gastro-duodenoscopy, ileo-colonoscopy and video capsule endoscopy) in the following 2 years. She is fit and asymptomatic at 5 years follow-up. PJS is an unusual autosomal dominant disease, characterised by gastrointestinal polyposis associated with mucocutaneous pigmentation and increased risk of gastrointestinal and extra-gastrointestinal malignancies. Chronic non-ischaemic intussusception could be the presenting feature of the PJS.^{1–3} Intussusception, on USS, may present the characteristic ‘target’ or ‘doughnut’ signs in cross-sectional images and the ‘telescope’ or ‘pseudo-kidney’ signs in longitudinal images. Many surveillance recommendation

Figure 1 (A and B) Upper gastrointestinal contrast study showing dilated duodenum and proximal jejunum with filling defect and mass effect in the proximal jejunum (arrows). (C–E) Ultrasound scan and colour Doppler study showing classic target/doughnut sign on cross-section and pseudo-kidney/telescope sign on longitudinal scan image and good vascularity on the colour Doppler study.

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strategies have been in place for gastrointestinal polyps/malignancies and other neoplasias.

Learning points

- ▶ Family history and perioral examination for mucocutaneous pigmentation may be rewarding in chronic abdominal pain and vomiting as our patient was labelled as abdominal migraine and cyclical vomiting.
- ▶ Ultrasound scan is diagnostic of intussusception and colour Doppler shows the vascularity and MRI scan or gastrointestinal contrast study or endoscopy with capsule video-endoscopy is helpful in finding remaining polyps.
- ▶ There are non-surgical options for diagnosis, reduction of intussusception and resection of the polyp should be considered whenever possible and feasible.

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Competing interests None.

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