Adult intussusception caused by Peutz–Jeghers hamartoma—an unusual presentation

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Abstract

A young woman presented with severe acute abdominal pain, superimposed on a past history of similar attacks. Ultrasonography revealed a small bowel intussusception. This was confirmed on laparotomy, and the lead lesion, an ileal polyp, was excised. Histology showed this to be a Peutz-Jeghers hamartoma. The possibility of intussusception should be kept in mind in adults presenting with acute abdominal pain. Ultrasonography can provide rapid and accurate corroboration.

Keywords

Adult; intussusception; Peutz-Jeghers; hamartoma; ultrasonography.
**Introduction**

Intestinal obstruction secondary to intussusception occurs very rarely in adults. It is a recognised feature of Peutz–Jeghers syndrome which tends to present at a younger age with characteristic circumoral pigmentation and multiple intestinal polyps. We report a case of intussusception in a young adult who presented with severe acute abdominal pain, without the other characteristic features of Peutz–Jeghers syndrome, but was subsequently diagnosed as having this condition.

**Case report**

A 33-year-old black Zimbabwean lady presented with a sudden onset of generalised abdominal pain that had progressively worsened for 24 h, associated with vomiting. She reported having suffered similar pains intermittently for the last 4 years and had been diagnosed as having irritable bowel syndrome. She had a long obstetric history but no relevant family history. On examination, she was pale and writhing around in extreme pain that could not be relieved by intravenous opioids and antispasmodics. There was no obvious pigmentation in the oral and buccal regions. Her abdomen was soft, distended and generally tender. She had a full rectum and active bowel sounds.

Her abdominal X-ray was unremarkable. An urgent ultrasound of her abdomen was organised, and this confirmed a small bowel intussusception. She was operated on immediately and an ileo-ileal intussusception was found with a large polyp as its lead (Fig. 1).

The intussusception was manually reduced and the lead lesion excised. She was discharged uneventfully 6 days later. Histological analysis of the polyp revealed it to be a Peutz–Jeghers hamartoma. The patient was referred for genetic screening.
Discussion

Adult intussusception is elusive and difficult to diagnose due to its vague presentation. Reported cases include a variety of non-specific and chronic symptoms such as abdominal pain, nausea and vomiting, recurrent bowel obstruction and fresh rectal bleeding. A review
of adults presenting with intussusception in Massachusetts found that they constitute 1% of all bowel obstructions and 5% of all intussusceptions (the remaining 95% being in children and juveniles)\[^1\]. Most were associated with a pathological lesion although a small number were idiopathic. Of the enteric lesions in this study, 48% were malignant and 52% were benign. In contrast, most colonic lesions were malignant.

Intussusception pre-dated by a long history of intermittent intestinal colic is a recognised feature of Peutz–Jeghers syndrome. This is an inherited autosomal dominant condition characterised by small bowel polyps and mucosal pigmentation usually found in the lips and buccal region. There is a 2–3% risk of malignant transformation which indicates the need for index cases to be referred to specialised units so that their families may be screened with endoscopy and genetic testing. The risk of malignancy is highest in stomach and small bowel lesions\[^1\].

**Learning points**

This case illustrates two learning points. The first is that intussusception should be considered as a possible diagnosis in adults who present with severe acute abdominal pain and a long history of intermittent bowel colic. Secondly, although computed tomography is said to be the investigation of choice, its availability may be restricted for a variety of reasons. Ultrasonography can be performed rapidly and accurately with minimum discomfort and can help make a quick diagnosis\[^2\]. The preferred treatment for adult intussusception is manual reduction and resection of the offending segment. In Peutz–Jeghers syndrome, a specialist referral ought to be made for further endoscopic and genetic tests.

**References**