Multiple Intussusceptions in Peutz-Jeghers Syndrome: Detection through Multidetector Computerized Tomography Enterography

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To the Editor: Peutz-Jeghers syndrome (PJS) is an autosomal dominant inherited disease, characterized by mucocutaneous pigmentation and hamartomatous gastrointestinal polyps. Due to the possible association between PJS and cancer, multidetector computed tomography (MDCT) enterography is regularly used for the surveillance of the small bowel to reduce polyp-related complications, particularly of intussusceptions. Given rare conditions of multiple intussusceptions in PJS, the study investigated a case of a 15-year-old girl who was diagnosed with concurrent intestinal intussusception and colon intussusception due to relatively large polyps by MDCT enterography. In this case, MDCT enterography was enabled to provide information of polyps and complicated intussusceptions in PJS for the consequent endoscopic enteroscopy or surgery.

The girl was sent to the outpatient department of Affiliated Hospital of Guizhou Medical University with an acute onset of dull pain at her right lower abdomen. On clinical examination and medical history inquiries, it was found that both patient and her mother had multiple lip and hand pigmentations. In addition, her mother had been gone through an abdominal surgery for adenocarcinoma in ileocecal junction 2 years ago. The patient completed CT enterography by MDCT scanners (Toshiba, Tokyo, Japan) with a section thickness of 0.5 mm. The coronal multiplanar reconstruction images [Figure 1a and 1b] and maximal intensity projection image [Figure 1c] revealed not only the site of intestinal intussusception and colon intussusception but also the inner loop of the bowel which was separated from the outer loop by a crescent of fat attenuation mesentery and intraluminal enhancing mesenteric vessels [arrow, Figure 1a and 1b]. Evidence of relatively large polyp was found adjacent to the intussusceptions [arrowheads, Figure 1a and 1b]. Meanwhile, multiple intraluminal polyps in jejunum, ileum, and colon were detected [arrowheads, Figure 1c]. The volume-rendered CT angiography image showed the supplied arteries of the polyps [arrowheads, Figure 1d]. Based on the above clinical and imaging characteristics, she was diagnosed with PJS with multiple intussusceptions. This was testified by upper gastrointestinal endoscopy and postoperative pathology. However, the patient died of postoperative gastrointestinal hemorrhage.

PJS is inherited, autosomal dominant disorder with an incidence from 1:25,000 to 1:280,000.[1] The accuracy of diagnosis is determined by the presence of histopathologically confirmed hamartomatous polyps and two following clinical criteria: family history, hyperpigmentation, and polyps in the small bowel.[2] PJS polyps occur anywhere in the gastrointestinal tract except the esophagus. They are most commonly seen in the jejunum, followed by the ileum. These polyps are frequently accompanied by recurrent abdominal pain due to intussusceptions whereas intussusceptions are often reduced spontaneously. Although conventional MDCT is useful in cases of acute abdominal pain resulting from intussusceptions, it might fail to identify small polyps of PJS. In such circumstances, MDCT enterography could be performed to detect small bowel tumors and complications in patients with PJS.[3] Our case proved that reconstruction images obtained from the isotropic data of MDCT could clearly display multiple polyps and two intussusceptions. The small intestine intussusception and colon intussusceptions were induced by the adjacent big polyps in our case study. Meanwhile, reconstruction images could be applied in observing the origin, blood supply, and size of PJS polyps. The diameter of polyps observed by MDCT enterography ranges from 5 mm to 30 mm. Most polyps are attached to intestinal wall with a wide or a narrow base, even a few have stems. It was also worth pointing out that the polyps in PJS should be

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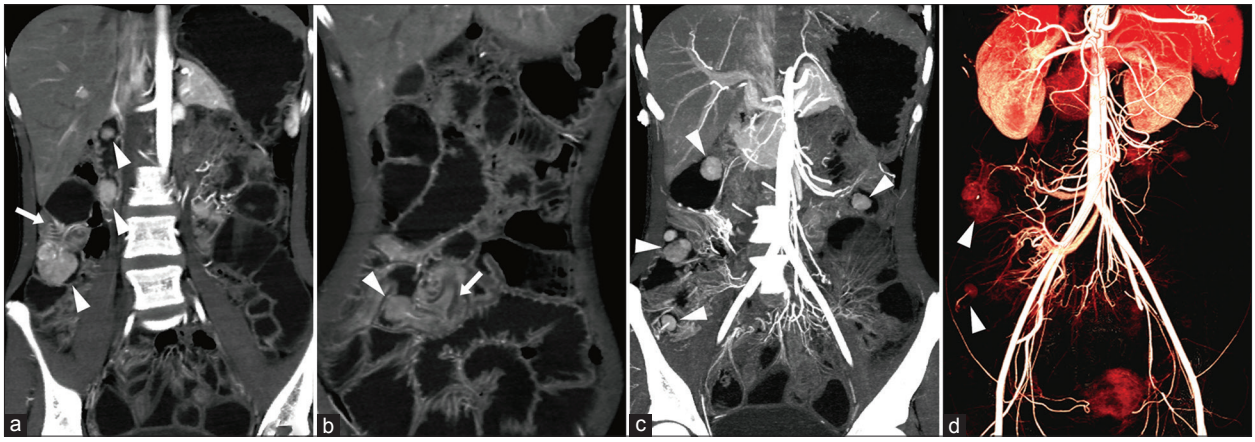

Figure 1: MDCT enterography of the patient (examined after administering 1350 ml oral contrast [water] containing 0.5% mannitol and 150 ml iohexol [300 mg/ml] intravenously at a rate of 4 ml/s). (a and b) MPR showed the intestinal intussusception and colon intussusception (arrows) and adjacent relatively large polyps (arrowheads). (c and d) MIP and CTA showed multiple intraluminal polyps and the supplied arteries of the polyps (arrowheads). MDCT: Multidetector computed tomography; MPR: Multiplanar reconstruction; MIP: Maximal intensity projection; CTA: Computed tomography angiography.

differentiated from juvenile polyposis, Cronkhite-Canada, and Bannayan-Riley-Ruvalcaba.

Our case proved that MDCT enterography was an efficient noninvasive and pain-free means of examination and a useful adjunct to capsule endoscopy in excluding strictures, clarifying ambiguous capsule endoscopic findings, or in searching for intramural or exoteric small bowel abnormalities.[4] It could provide information of polyps and complicated intussusceptions in PJS for the consequent endoscopic enteroscopy or surgery.

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Conflicts of interest
There are no conflicts of interest.

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