Sonographic Findings of Intussusception in Peutz-Jeghers Syndrome — A Case Report and Review of the Literature

Yih-Shing Yao, Wei-Hong Chen*, Ying-Ho Cheng

Peutz-Jeghers syndrome is a rare hereditary disorder characterized by polyps in the gastrointestinal tract and focal mucocutaneous pigmentation. Some patients also suffer from intussusception. Early diagnosis of intussusception in patients with Peutz-Jeghers syndrome is important. With clear past history taking and sonographic examination, an exact diagnosis can be made preoperatively. Here, we present a patient with Peutz-Jeghers syndrome in whom sonography was used to detect intussusception when he presented with acute abdomen. The symptoms were relieved by surgical treatment.

KEY WORDS — intussusception, Peutz-Jeghers syndrome

Introduction

Peutz-Jeghers syndrome (PJS) is an autosomal-dominant disorder characterized by typical mucocutaneous pigmentation and gastrointestinal hamartomatous polyps [1–3]. Polyps in PJS may cause intussusception, although the incidence with which this occurs is unknown. Early diagnosis of intussusception is important because it may be treated conservatively. We present a patient with PJS in whom sonography was used to detect intussusception that was successfully treated by surgery.

Case Report

A 20-year-old man had suffered from intermittent abdominal pain that was relieved spontaneously for 3 years. He visited our emergency room due to abdominal pain of 2 days’ duration without improvement after medical treatment by a local medical doctor. Physical examination showed diffuse abdominal tenderness and rebounding pain. Some skin pigmentation was seen in the perioral area, fingers and toes (Fig. 1). Abdominal sonography showed intussusception in the left upper abdomen. Scanning of the affected bowel showed polyps in the central echogenic area (Fig. 2). PJS with polyp-induced intussusception was diagnosed.

Emergent laparotomy showed a 30 cm segment of jejunum with intussusception. Bowel resection was performed due to ischemic change and a polyp measuring 4 cm in size was removed (Fig. 3). Pathology showed PJS with hamartomatous change (Fig. 4). The patient recovered smoothly and was discharged 1 week after operation.

Department of Surgery, Cardinal Tien Hospital Yung-Ho Branch, Taipei, Taiwan, R.O.C.

*Address correspondence to: Dr. Wei-Hong Chen, Department of Surgery, Cardinal Tien Hospital Yung-Ho Branch, 80 Chung-Hsing Street, Yungho, Taipei 234, Taiwan, R.O.C. E-mail: t00800.yao@msa.hinet.net
PJ(S) is a rare autosomal-dominant inherited disorder that is characterized by mucocutaneous pigmentation and hamartomatous polyps in the gastrointestinal tract, predominantly in the jejunum [1–3]. In a Japanese series of 222 patients with PJ(S), the average age at diagnosis in males was 22 years.

**Fig. 1.** Multiple mucocutaneous pigmentation.

**Fig. 2.** Ultrasound shows bowel wall thickening and polyps within the bowel. Arrows indicate the incarcerated polyp.

**Discussion**

PJ(S) is a rare autosomal-dominant inherited disorder that is characterized by mucocutaneous pigmentation and hamartomatous polyps in the gastrointestinal tract, predominantly in the jejunum [1–3]. In a Japanese series of 222 patients with PJ(S), the average age at diagnosis in males was 22 years.
intussusception may show a mass with multiple concentric rings [6,7]. In this patient, the hypoechoic layers represented the bowel wall, while the polyp presented as a central echogenic area (Fig. 4). Color Doppler ultrasound may show many blood flow signals in the intussuscepted bowel wall [7]; some authors suggest that it may reflect the degree of bowel ischemia. Computed tomography can also be used, and it may show a mass with a target appearance [7]. In conclusion, we reported a case of PJS in which sonography was used for the actual diagnosis. Ultrasound is a good preoperative diagnostic tool for PJS with intussusception.

References