Role of capsule endoscopy in blue rubber bleb nevus syndrome

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The blue rubber bleb nevus syndrome (BRBNS) is a rare disease characterized by skin lesions and intestinal venous malformations that lead to gastrointestinal bleeding and chronic anemia. About 200 cases have been published. Surgery is indicated for most of the persistent bleeders.[1] Effective treatment of the venous malformations includes endoscopic ablation and surgery. Video capsule endoscopy (VCE) is a new, noninvasive imaging technique that can be used for the whole small bowel.[2] We report a case of BRBNS detected with VCE in which the patient had had numerous small-bowel lesions since childhood.

Case report

A 35-year-old man was admitted with melena and chronic anemia. He had a history of several hospital admissions for lower gastrointestinal bleeding, which was treated conservatively with iron supplements and whole blood transfusions of more than 5 units a year over 30 years. On physical examination, we found dark blue, soft, lobulated lesions (5 mm–1 cm in size) on his tongue, shoulders and extremities. Mucous surfaces were free of bleb-like lesions. Laboratory tests revealed iron deficiency and anemia. His hemoglobin level was 70 g/L, with a mean corpuscular volume of 69 fL. On peripheral blood smear, the red blood cells were microcytic and hypochromic, indicating iron deficiency. Other laboratory tests and viral serology findings were within normal limits. Colonoscopy revealed 2 papular venous malformations, 1 cm in size, at the sigmoid colon and the hepatic flexure. To search for additional lesions, we carried out enterocolysis and identified multiple pedunculated filling defects in the jejunum and ileum. We instructed the patient to swallow a VCE device, and the following computer analysis demonstrated multiple pink-bluish pedunculated and sessile venous malformations 1–2 cm in diameter (Fig. 1). Since we could not prevent blood loss using endoscopic control and conservative therapy, we recommended surgical ablation of the lesions.

At laparotomy, we found venous malformations located in various parts of the small bowel. In addition to 2 different lesion-guided invaginations, we were able to see all the lesions from the external surface of the bowel. For 9 lesions larger than 1 cm, we performed ligation-excision through an enterotomy (Fig. 2); for 8 lesions that were smaller than 1 cm, we adopted a pursestring-suture occlusion technique without an enterotomy.

Histopathologic examination of the lesions revealed blood-filled ectatic vessels, lined by a single layer of endothelial cells with surrounding thin connective tissue.

The man's postoperative course was uncomplicated with the use of short-term somatostatin therapy, and he had a normal gastrointestinal passage within 1 week of surgery. During follow-up, he had neither occult blood in his feces nor anemia. Two years postoperatively, he was well and free of all medication.

Discussion

Blue rubber bleb nevus syndrome consists of bluish venous malformations of the gastrointestinal tract, skin and other organs. The abdominal location of nevi has been reported in solid viscera (liver, spleen, kidney) and in the digestive tract either in a diffused or segmental form.

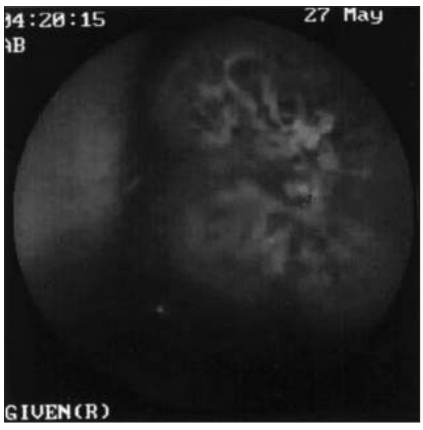

FIG. 1. Typical view obtained by video capsule endoscopy.

FIG. 2. Ligation and excision of a lesion through an enterotomy.

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A familial form has been described with an exclusively male pattern, but no genetic basis has been clearly demonstrated. Blue rubber bleb nevus syndrome nodules contain dysplastic venous channels with flat endothelium and irregularly attenuated walls with deficient smooth-muscle cells.[3] Bleeding occurs frequently from the gastrointestinal tract malformations and can result in occult blood loss and iron deficiency anemia. There may be abdominal pain, intussusception, volvulus, infarction or internal hemorrhage.

The diagnosis of BRBNS is initially made from the clinical cutaneous findings of characteristic lesions and on a stool guaiac test to detect anemia and gastrointestinal bleeding. Evaluation of the gastrointestinal tract to identify bleeding sites may be accomplished with barium studies, angiography, tomography, magnetic resonance imaging and endoscopy.[4] Video capsule endoscopy provides good visualization of gastrointestinal mucosa and helped us detect BRBNS and its extension in the small bowel. The diagnostic yield from VCE appears to be superior to small-bowel radiography and push enteroscopy.[2]

The treatment of gastrointestinal venous malformations depends on the symptoms: a conservative approach is usually recommended when the clinical features and bleeding episodes are mild. Although there are a few reports of medical therapy such as corticosteroids and interferons for venous malformations, sustained response has not been reported.[5] Since the lesions are usually transmural, safe endoscopic eradication is not possible because of a high risk of perforation. Surgical treatment with and without intraoperative endoscopy is the best approach for persistent bleeders. We employed resection through an enterotomy and extramural occlusion with a pursestring suture technique in our patient. Because new lesions may form postoperatively, the surgeon must choose minimal resection procedures.

Competing interests: None declared.

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