^{99m}Tc-Labeled RBC Scan in a Patient with Blue Rubber Bleb Nevus Syndrome: A Case Report

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Abstract Blue rubber bleb nevus syndrome (BRBNS) is a rare disorder characterized by cutaneous and gastrointestinal vascular malformations. We report here the case of a 13-year-old patient with BRBNS who was suffering from chronic anemia. His chief complaint was melena. Contrast-enhanced abdominal computed tomography showed multiple hemangiomas in the small bowel. ^{99m}Tc-labeled red blood cell (RBC) scan identified the bleeding sites in the small bowel. The patient underwent exploratory laparotomy with segmental resection and multiple wedge resection of the small bowel. The pathological results confirmed the multiple cavernous hemangiomas. This case confirms the critical role of the ^{99m}Tc-labeled RBC scan in localizing the bleeding sites in patients with BRBNS.

Keywords Blue rubber bleb nevus syndrome · ^{99m}Tc-labeled red blood cell scan · Venous malformation · Chronic anemia

Introduction

Blue rubber bleb nevus syndrome (BRBNS) is a rare disorder characterized by multiple cutaneous venous malformations in the skin and gastrointestinal (GI) tract. The venous malformations in the GI tract are associated with intestinal hemorrhage and result in iron deficiency anemia. The disease was first reported in 1860 as the association between hemangiomas in the skin and in the GI tract [1]. It was called “blue rubber bleb nevus syndrome” in 1958 [2]. The vascular

malformations in BRBNS are mainly composed of cavernous hemangiomas [3]. They may involve the brain, liver, lung or muscle [4]. Patients usually exhibit GI bleeding at an early age that continues throughout their life. Sudden massive hemorrhage rarely occurs. We report the case of a patient with BRBNS with GI bleeding in whom the bleeding sites were imaged by ^{99m}Tc-labeled red blood cell (RBC) scanning.

Case Report

A 13-year-old boy presented to the emergency room of Seoul National University Bundang Hospital on October 2012. His chief complaint was melena. He also had symptoms of intermittent dizziness, exertional dyspnea and headache that had initially developed 8 months previously.

He had no medical history of nonsteroidal antiinflammatory drug use or peptic ulcer. He had attended the hospital due to a pale complexion and recurrent abdominal pain 3 years previously and had been followed up. At that time, iron deficiency anemia was diagnosed and intestinal hemangioma and liver hemangioma were detected by abdominal sonography. His intermittent abdominal pain was thought to have originated from recurrent intussusception due to the intestinal hemangioma. There was no evidence of active GI bleeding. Accordingly, he received iron replacement therapy without surgery.

Since infancy, he had had multiple cutaneous hemangiomas on the chest, abdomen, hands, feet and popliteal areas. The size of the hemangiomas was less than 5 mm. He had three surgical operations on a hemangioma of the right index finger. However, he had no family history of any skin lesions or recurrent GI bleeding.

On physical examination, he was very pale. Some small hemangiomas were observed on a finger, both lower extremities and the trunk (Fig. 1). His hemoglobin level was 6.9 g/dL, hematocrit 23.8 %, mean corpuscular volume

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Fig. 1 Subcutaneous venous malformations in a 13-year-old boy with blue rubber bleb nevus syndrome. **a** Deformity of the index finger of the right hand. **b** The plantar surface of the left toe, the popliteal fossa and the trunk show subcutaneous venous malformations

60.5 fL and mean corpuscular hemoglobin concentration 28.8 g/dL. Serum iron was 254 $\mu\text{g/dL}$, ferritin <3 ng/mL and total iron-binding capacity 495 $\mu\text{g/dL}$. The blood test showed iron-deficiency anemia. The hemoglobin level was lower than in the outpatient clinic (9.9 g/dL). On the basis of these results, transfusion of packed red blood cells was started. After hospitalization, there was no melena.

The contrast-enhanced abdominal CT scan showed several small enhancing lesions in the small bowel (Fig. 2). Strongly enhancing lesions are characteristic of vascular lesions suggesting hemangioma. Endoscopic evaluation was not performed. In order to detect the bleeding focus, a $^{99\text{m}}\text{Tc}$ -labeled RBC scan was performed. Sequential images of the abdomen and pelvis were obtained over 3 h. The scan showed two bleeding sites in the pelvic cavity and the left

lower abdominal cavity after 40 min (Fig. 3). Active bleeding from the small bowel in the pelvic cavity was identified on sequential images. The patient underwent exploratory laparotomy with segmental resection and multiple wedge resection of the small bowel. Numerous hemangiomas were located throughout the small bowel (Fig. 4), liver and peritoneum. A total of eight hemangiomas were removed. Multiple lesions with less than 5 mm in size were not removed due to low probability of bleeding. A small-bowel intussusception caused by a hemangioma was observed above 30 cm from the ileocecal valve, and manual reduction was performed. This lesion was thought to be the cause of the abdominal pain. A limited bowel resection was performed.

A pathological specimen confirmed the dilated cavernous blood-filled spaces in the small intestinal wall, consistent with cavernous hemangioma (Fig. 5).

The patient was discharged from hospital on the 5th day after operation. After 10 days, he attended the outpatient clinic of our hospital. He denied melena and abdominal pain.

Discussion

BRBNS is a rare disorder characterized by multiple vascular malformations [3]. The lesions of BRBNS are typically small, circumscribed, and multifocal. They may be present in any body tissue. Affected tissues and organs include the skin, subcutaneous tissue, muscle, bone, joint, parotid gland, thymus, lung, liver, gallbladder, pancreas, spleen, mesentery, pelvis, and retroperitoneum. However, the predominant organs affected are the skin and GI tract. Although apparently autosomal dominant transmission has been reported in some patients [5], most cases are sporadic.

Fig. 2 Contrast-enhanced abdominal CT images show multiple hemangiomas (arrows) in the small bowel suggesting venous malformations

Fig. 3 ^{99m}Tc -labeled RBC images show two bleeding sites (arrows) in the left lower abdominal cavity and the pelvic cavity after 40 min. The sequential images at 150 min, 160 min, 170 min and 180 min identified ^{99m}Tc -labeled RBC movement through the bowel in the abdominal cavity, which represents bleeding of the small bowel

The skin lesions in BRBNS usually do not require treatment except for cosmetic purposes. In such patients, treatment usually consists of laser therapy [6]. However, repeated treatments are often necessary because of recurrence. Larger soft-tissue lesions, particularly those that are symptomatic, can be treated by direct percutaneous sclerotherapy using substances such as ethanol and sodium tetradecyl sulfate [4]. Lesions in the esophagus, stomach, proximal duodenum and colon may be treated with endoscopic band ligation or sclerotherapy [4, 6]. GI lesions may clinically manifest with bleeding or may be a lead point for small-bowel intussusceptions [7, 8]. The bleeding in the GI tract is typically chronic continual bleeding rather than sudden catastrophic hemorrhage. Most patients have a history of chronic anemia

in childhood and BRBNS is diagnosed by GI bleeding in adolescence [9].

Small-bowel lesions require surgical resection or ligation [7]. Some studies have investigated medical therapy using steroids or interferon α -2a to control the chronic blood loss in patients with BRBNS [10], but have shown no definite benefit. This result is not unexpected because lesions in BRBNS are venous malformations, not vascular neoplasms, and thus would not be expected to respond to angiogenesis inhibitors [4]. Surgical excision has been discouraged because of the multiple lesions and the belief that resected lesions will recur.

Fishman et al. reported their clinical experience in ten surgically treated patients with BRBNS [7]. Lesions were

Fig. 4 Intraoperative photograph shows two small-bowel malformations

Fig. 5 A histological section of a cavernous hemangioma from the small intestine

identified using complete GI endoscopy. The lesions were distributed throughout the GI tract in all sections from mouth to anus, but predominated in the small and large bowel. The multiple venous malformations were removed by a combination of segmental bowel resection, wedge resection, suture ligation, polypectomy, and band ligation. A mean of 137 focal GI venous malformations per patient were resected. They eliminated the bleeding in nine of ten patients over a follow-up period of 5 years. In one patient the small bowel was not evaluated. They suggested that serious chronic bleeding from GI venous malformations of BRBNS can be controlled by aggressive resection of the malformations, regardless of their number and location.

We present here the case of a patient with BRBNS. This patient was suffering from chronic iron deficiency anemia and intermittent abdominal pain. He was admitted to our hospital because of melena. The enhanced CT scan identified several hemangiomas of the small bowel and a ^{99m}Tc -RBC scan showed active bleeding of the small bowel. At laparotomy, multiple hemangiomas were found throughout the small bowel. Surgical specimens confirmed cavernous hemangiomas.

Conflict of Interest We declare that we have no conflict of interest.

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