

Blue Rubber Bleb Nevus Syndrome: Small Bowel Involvement and Wireless Capsule Endoscopy Utility

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ABSTRACT

A 27-year-old woman presented with anemic symptom for 2 months but no obvious gastrointestinal bleeding was detected. She is a known case of blue rubber bleb nevus syndrome (BRBNS). She previously underwent multiple band ligation and sclerotherapy because of multiple vascular lesions in her stomach, small bowel, colon since 14 years ago. Capsule endoscopy revealed many purplish blebs in her entire small bowel. Some of them were bleeding and blood-stain blebs. According to our knowledge, it is the first case report of capsule endoscopic picture of BRBNS in the small bowel.

Key words : Blue Rubber Bleb Nevus Syndrome, capsule endoscopy

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INTRODUCTION

Blue rubber bleb nevus syndrome (BRBNS) is a rare syndrome characterized by vascular anomalies in GI tract and cutaneous hemangioma. A familial history is infrequent, although a few cases of autosomal dominant transmission have been reported⁽¹⁾. The cutaneous hemangioma can be found on the trunk, extremities, and face. Small bowel is the most common site for GI tract involvement⁽²⁾. According to our knowledge, this is the first case report of capsule endoscopic picture of BRBNS in small bowel.

CASE REPORT

A 27-year-old woman presented with anemic symptoms without hematemesis or melena for 2 months duration. Fourteen years ago, she presented with hematemesis and melena and physical examination revealed violaceous, non pulsatile cutaneous lesions on her hand, shoulder, dorsum, mouth and feet. Upper endoscopy and colonoscopy disclosed multiple purplish polypoid lesions varying in size in the stomach and colon. Hemostasis was achieved by a combination of absolute alcohol injection into blebs and band ligation.

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All cutaneous lesions were removed by either surgical excision or pulse dye laser later. BRBNS was diagnosed since then. Over the last 13 years, she underwent upper and lower endoscopy with sclerotherapy and band ligation for several times. Her latest colonoscopy 2 years ago revealed normal. She felt well since then until this presentation. There was no history of non steroidal anti-inflammatory drug (NSAIDs) use, peptic ulcer, or chronic liver disease. Physical examination revealed stable vital sign, pale conjunctiva. Neither abnormal skin lesion nor mucosal lesion were found by physical examination.

Hemoglobin on admission was 7.8 gm/dL (normal 12.5-15.5 gm/dL). White blood count and differential were within normal limit. Heme occult blood showed a positive result. Other blood tests were un-

remarkable. Capsule endoscopy was planned in this patient because of a possibility of small bowel lesions. The result of capsule endoscopy revealed significant findings. There were 1 purplish polypoid bleb in the stomach (Figure 1) and more lesions varying in size from 0.5-2 cm. in the small bowel (Figure 2, 3). Some of these lesions showed blood-stained bleb and bleeding which also detected by SBI function of Given Imaging System (function which will catch red color from motion picture taken from capsule endoscopy) as shown in Figure 4.

DISCUSSION

BRBNS (Bean's disease) was first reported in 1860 by Gascoyen GG3 which described an associa-



Figure 1 A purplish polypoid bleb in the stomach.



Figure 3 Potentially bleeding lesion in small bowel.



Figure 2 Another small bowel lesion.



Figure 4 Blood-stained bleb and bleeding.

tion between cavernous hemangiomas of the skin and similar lesions in the GI tract. In 1958, Bean further described these lesions and called the term blue rubber bleb nevus syndrome⁽⁴⁾. BRBNS is a rare disorder which most cases are sporadic, but autosomal dominant inheritance has been reported. The disease tends to appear mainly in childhood, but a few adult cases have been described⁽⁵⁾. Although the cutaneous and GI systems most frequently are involved with these lesions but the central nervous system, eyes, thyroid, parotid gland, oral cavity, lungs, kidney, liver, spleen, musculoskeletal and bladder also be affected.

The lesion is typically discrete purplish, bluish vascular lesion which may be flat, macular, or polypoid⁽⁶⁾. Blood-filled ectatic vessels, lined by a single layer of endothelial cells, with surrounding thin connective tissue are the characteristic pathology.

Most patients commonly presented with asymptomatic skin lesions and have a normal life span without any malignant transformation. Extent of visceral organ involvement is one of the major prognostic factors. Some patients may have fatal GI bleeding and rarely, central nervous system involvement can be fatal. Bones and joints lesions are also able to cause some functional and physical morbidity, requiring amputations in some cases. For patients with BRBNS whose presenting symptom was abdominal pain, intussusception, volvulus, and bowel infarction should be considered.

For GI involvement, BRBNS is an uncommon cause of GI bleeding. In oppositely, the GI bleeding is the most common mode of its presentation⁽⁷⁾. Most of GI bleeding is slow, minor, chronic, and occult, resulting in iron deficiency anemia from ongoing loss. Lesions may involve any portion from oral to anal mucosa but are most common in the small bowel and distal large bowel⁽⁶⁾. Previously, they are best seen by enteroscopy and not well visualized by other imaging technique. Nowadays, wireless capsule endoscopy is becoming popular for obscure GI bleeding. We herein

report the small bowel pictures of BRBNS detected by capsule endoscopy. The 8-fold magnification from capsule endoscopy compare to regular endoscopy has a significant impact on clarification of picture.

In addition, the capsule can detect more lesions mainly in the area that can not be reached by a standard enteroscope. Therefore, patients with BRBNS may need capsule endoscopy prior to definite treatment such as surgery or therapeutic enteroscopy since there is a possibility of more lesions beyond routine enteroscopy techniques. Sometimes, new GI lesions may continue to occur, hence patients need periodic GI and hematologic surveillance studies. Capsule endoscopy, in our opinion, is potentially going to be the best screening tool because of their advantages of less invasive and 8-fold magnification. Finally, it is a total small bowel length examination.

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