Surgery for Blue Rubber Bleb Nevus Syndrome—a Case Report

U. Aravindan¹ · R. Ganesan² · M. Thamarai Kannan³

Received: 21 April 2017 / Accepted: 8 December 2017 / Published online: 19 December 2017

© Association of Surgeons of India 2017

Abstract

Blue rubber bleb nevus syndrome (BRBNS) is a rare disease characterised by multiple venous malformations and haemangioma in the skin and visceral organs. The lesion often involves the cutaneous and gastrointestinal tract. Most common visceral organ affected is the GIT. Most predominant region involved in the GIT is small bowel. However, vascular lesions can occur anywhere from oral mucosa to anal canal. GIT bleed is relatively slow, resulting in minor, chronic and occult blood loss. The syndrome in the GIT may also present with severe complications such as rupture, volvulus, intussusceptions and even death. Cutaneous malformations are usually asymptomatic and do not require treatment. The treatment of GIT lesions is determined by the extent of intestinal involvement and severity of the disease. Most patients respond to supportive therapy such as iron supplementation and blood transfusion. Surgical resection, endoscopic sclerosis and laser photocoagulation have been proposed for more significant haemorrhage and severe complications. Here, we present a case of BRBNS in a 13-year-old girl involving the GIT especially the large bowel, presenting with the complaints of bleeding per rectum and iron deficiency anaemia. Initially, endoscopic sclerotherapy was performed, but to no response. Hence, proceeded with surgical resection.

Keywords Blue rubber bleb nevus syndrome · Haemangioma · Vascular malformations · Gastrointestinal bleeding

Introduction

Blue rubber bleb nevus syndrome (BRBNS) is a rare congenital disorder associated with multiple venous malformations in the skin and visceral organs. BRBNS was initially discovered in 1860 by Gascoyen. Later, it was made famous in 1958 by William Bennett Bean who coined the term BRBNS, followed by which the disease has been termed bean syndrome. A MEDLINE search yielded about 200 case reports published to date.

Case Report

A 13-year-old girl presented with bleeding per rectum and fatigue for 2-month duration. The patient had no significant family history. Physical examination revealed pallor and no skin lesions. The abdomen examination was normal. Digital rectal examination showed blood-stained faecal staining. Other system examinations are normal. Laboratory findings were as follows: Hb—3.8 g/dl; RBC—2.7million cells/mm³; WBC—6900 cells/mm³; DC—P78%, L18% and M2%; platelet count—2.8 lakhs; PCV—14%; MCV—12 pg and mean corpuscular haemoglobin concentration—25.6 g/dl, RBS—90 mg/dl, urea—28 mg/dl and creatinine—0.9 mg/dl. Peripheral smear showed RBC having cells with microcytic hypochromic and moderate anisopoikilocytosis, and WBC being normal in count. Few neutrophils show toxic vacuolation, platelet—adequate, BT—2 min, CT—4 min and 20 s, USG abdomen and pelvis—normal study, UGI scopy—normal study and colonoscopy—blue rubber bleb lesions noted in the rectum, sigmoid colon and descending colon (Fig. 1). Rest of the colon did not show these abnormalities. On the basis of the above said findings of venous malformations, the diagnosis was compatible with BRBNS. Patient was treated with iron supplementations along with blood transfusions. Endoscopic

✉ U. Aravindan

R. Ganesan
ganesantmch@gmail.com

M. Thamarai Kannan
sushma.88s@gmail.com

¹ Department of SGE, TMCH, Thanjavur, India

² Department of MGE, TMCH, Thanjavur, India

³ Department of General Surgery, TMCH, Thanjavur, India

Fig. 1 Colonoscopy image showing blue bleb lesion in descending colon

sclerotherapy was given for two sittings at 1-month interval. The sclerosant used was 1% polidocanol. She was managed conservatively. The patient improved well after 3 days. The patient was followed up for a year. Again, she presented with hematochezia and fatigue associated with occasional melena. The patient got readmitted and further investigated. In view of recurrent episodes of hematochezia, blood picture showing iron deficiency anaemia requiring multiple blood transfusions and failure of endoscopic management, surgery was planned.

Procedure

The abdomen was entered through midline incision. Perioperatively, clusters of lesions were found involving the rectum, sigmoid and descending colon. Few lesions were found scattered over the transverse and ascending colon. The lesions were seen in the serosal aspect. Total proctocolectomy with ileoanal J-pouch anastomosis along with loop ileostomy was done (Fig. 2).

The postoperative period was uneventful. She has been completely asymptomatic till date. Histopathology examination showed many dilated vascular channels filled with blood in the colonic mucosa and submucosa. The muscularis propria appears normal. Serosal congestion was present.

Ileostomy was closed after 3 months. Her condition is good.

Fig. 2 Intraoperative findings showing scattered hemangioma lesion in the sigmoid and descending colon with cut section showing blue rubber lesion

Discussion

BRBNS incidence rate is 1:14,000 [1]. It affects all races, most common among the Caucasians. The cause of this syndrome is unknown. Indian case reports are few in number and most of them are from North India. There have been two published reports from South India [2, 3]. The case reports from India all are sporadic and mostly in males. Most are associated with skeletal deformities in addition to cutaneous and GI vascular malformation. Few case reports show involvement of GIT alone [4].

The diagnosis of BRBNS is based on the presence of characteristic cutaneous lesions with or without GI bleeding and/or the involvement of other organs. Cutaneous lesions present as multiple protuberant, dark blue, compressible blebs, a few millimetres to several centimetres in diameter and can vary in morphological shape. Our patient had iron deficiency anaemia and hemangioma in the large intestine.

For GI lesions, the diagnostic workup includes UGI scopy, CT and MRI abdomen. Colonoscopy is the ideal exam for the diagnosis of the venous malformations located in the colon. Video capsule endoscopy is a new, non-invasive technique used for diagnosing the GIT lesions. Selective arteriography and scintigraphy are useful in identifying the source of the bleeding [5].

The treatment of GIT BRBNS is determined by the extent of intestinal involvement and severity of the disease. If bleeding is minor or intermittent, iron supplementation and blood transfusion is done. Endoscopic removal is the effective modality treatment for repeated bleeding. Other therapeutic modalities include bipolar or argon plasma coagulation, snare resection, endoscopic sclerotherapy, rubber band ligation and polypectomy [6]. A total colectomy or segmental resection of the small intestine is reserved for cases where the disease is extremely localised or in cases of bleeding refractory to conservative measures [8]. Fishman et al. showed their experience with complete and extensive surgical resection in ten patients with excellent results [9], though only nine reports exist in

literature involving extensive surgical resection for these gastrointestinal vascular lesions.

In our case, the colon is involved, which is a rare presentation, and no skin or other organ involvement is seen. So, total proctocolectomy with ileoanal pouch anastomosis with loop ileostomy was done in this case, since the patient had recurrent episodes of GI bleeding and failed endoscopic sclerosant therapy management.

Conclusion

The GIT lesions of BRBNS are more clinically relevant than the skin and soft tissue lesions. Though the surgery may be demanding and tedious, a good multidisciplinary approach with adherence to standard surgical principles results in successful outcome with minimal morbidity.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

References

1. Gentry R, Dockerty MB, Clagett OT (1949) Vascular malformations and vascular tumours of the gastrointestinal tract. *Int Abst Surg* 88: 281–323
2. Krishnappa A, Padmini J (2010) Blue rubber bleb nevus syndrome. *Indian J Pathology Microbiology* 53:68–170
3. Manoj kumar Sahu, Ebby George Simon, Joseph Anjilivelil Joseph, Ashok Chacko Dept. of GI Sciences, Christian Medical College, Vellore. BRBNS: case report and review of literature 2010; 1(4): 183–185
4. Teixeira MG et al (2003) Blue rubber bleb nevus syndrome: case report. *Rev. Hosp. Clin. Fac. Med. S Paulo* 58(2):109–112
5. Huo JR, Zhang J, Wang XH, Liu DL, Ou DL, Liu XW (2005) Clinical and endoscopic characteristics in blue rubber bleb nevus syndrome. *Chinese J Digestive Endoscopy* 22:194–195
6. Apak H, Celkan T, Ozkan A et al (2004) Blue rubber bleb nevus syndrome associated with consumption coagulopathy treatment with interferon. *Dermatology* 208:345–348
8. Choi KK, Kim JY, Kim MJ, Park H, Choi DW, Choi SH, Heo JS (2012) Radical resection of intestinal blue rubber bleb nevus syndrome. *J Korean Surgical Society* 83(4):316–320. <https://doi.org/10.4174/jkss.2012.83.5.316>
9. Fishman SJ, Smithers CJ, Folkman J, Lund DP, Burrows PE, Mulliken JB, Fox VL (2005) Blue rubber bleb nevus syndrome: surgical eradication of gastrointestinal bleeding. *Ann Surg* 241(3): 523–528. <https://doi.org/10.1097/01.sla.0000154689.85629.93>