Annals of Medicine and Surgery 5 (2016) 93–96

Contents lists available at ScienceDirect

Annals of Medicine and Surgery

journal homepage: www.annalsjournal.com

The surgery for blue rubber bleb nevus syndrome

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HIGHLIGHTS

- Our results indicated the endoscopy examination was the best diagnostic way for Blue Rubber Bleb Nevus Syndrome.
- The surgical eradication was the most effective treatment for Blue Rubber Bleb Nevus Syndrome.
- Patients with an overwhelming number of lesions may require staged operative procedures.

ARTICLE INFO

Article history:
Received 21 September 2015
Received in revised form
28 December 2015
Accepted 28 December 2015

Keywords:
Blue rubber bleb nevus syndrome (BRBNS)
Venous malformations (VM)
Surgery

ABSTRACT

Blue rubber bleb nevus syndrome (BRBNS) is a rare vascular anomaly syndrome consisting of multifocal venous malformations (VM). The malformations are most.

prominent in the skin, soft tissues and gastrointestinal (GI) tract. Chronic bleeding induced by the vascular malformation (VM) lesions in gastrointestinal (GI) tract was the main clinical problem which should be treated. In this paper, two patients with BRBNS were treated by surgery for gastrointestinal bleeding. Our results indicated the endoscope examination was the best diagnostic way for VM in GI. Although many therapeutic methods have been tried, aggressive surgical eradication was the most effective for venous anomalies that cause GI bleeding in BRBNS.

1. Introduction

Blue rubber bleb nevus syndrome (BRBNS) is a rare syndrome characterized by numerous venous malformations (VM) which can be found in any tissue or organs [1–3], but most common in the skin and gastrointestinal (GI) tract [4–6] with a diameter of 1–2 cm and blue color appearance. The patients with BRBNS usually present serious anemia throughout their life because of chronic GI blood loss. Two BRBNS patients with GI bleeding were treated with surgery and surgical experiences were discussed (The informed consent was obtained from the patients).

2. Materials and methods

2.1. Case 1

Female, 14 years old, repeated GI bleeding for 7 years without family history. Bleeding could not be improved by blood transfusion. Multi-hemangiomas appeared in her skin at the early age. Hemangioma excision of left popliteal fossa had been performed at 7-year-old.Gastric multi-polyps were shown by gastroscopy. Hepatic hemangioma and splenomegaly were found in ultrasonography. DSA indicated a number of small nodular staining in hepatic parenchymal and small abnormal staining with irregular and boundary ambiguity in jejunum artery, suggesting the possibility of hemangioma or malformation.

In laparotomy, several hemangioma-like lesions were found in the surface of liver. A segment of ileum including 14 lesions was resected, other 3 lesions were removed separately, and the lesions of liver surface were ligated.

One year after surgery, patient had a follow-up with satisfactory outcomes: black stool disappeared and hemoglobin maintained at

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http://dx.doi.org/10.1016/j.amsu.2015.12.062

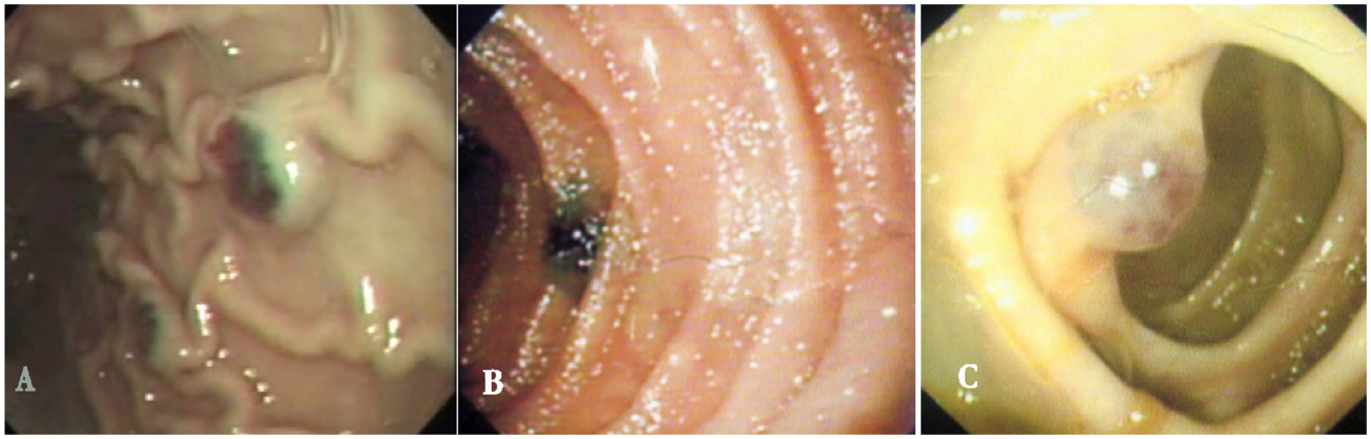

Fig. 1. Endoscopy showed numerous lesions in GI. A:Stomach; B:Duodenum; C:Colon.

more than 120 g/L without additional iron supplement and blood transfusion.

2.2. Case 2

Female, 10-years-old, without family history, GI bleeding firstly occurred at 5-year-old, then gradually frequent and severe. Patient also had a history of skin hemangioma excision. Iron replacement therapy had been given since 7-years-old without any improvement. Weekly blood transfusion were required, but hemoglobin only been maintained at 40–50 g/L.

In physical examination, several scattered blue lesions with different size, which were easily ruptured, were found in limbs (Fig. 2A). Endoscopy showed that the purple nodular lesions were widely distributed over the stomach, duodenum and upper jejunum. Colonoscopy also indicated multiple lesions in colon

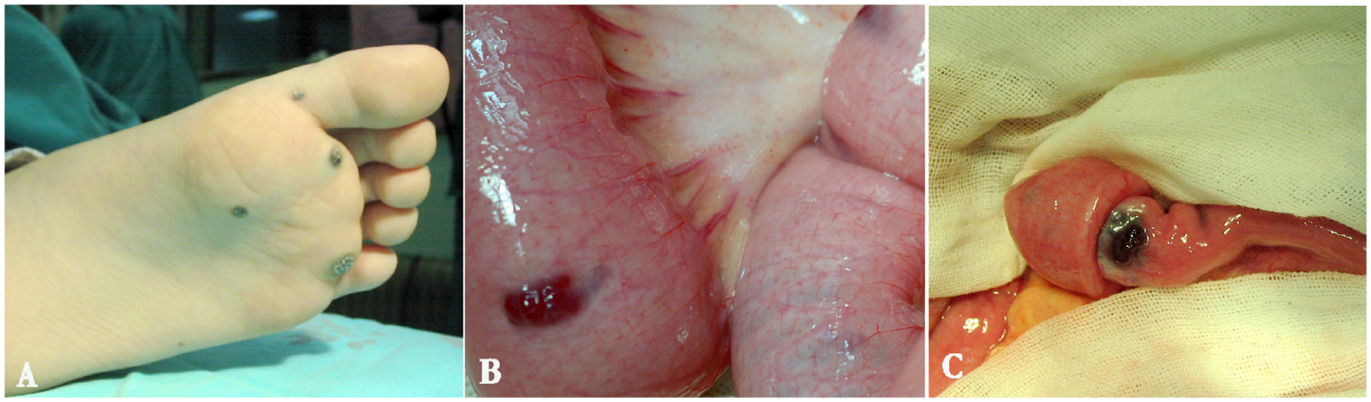

Fig. 2. A: Blue lesions with different size at the foot. B: Trans-wall lesions at the serosa side. C: Ileum intussusception induced by VM.

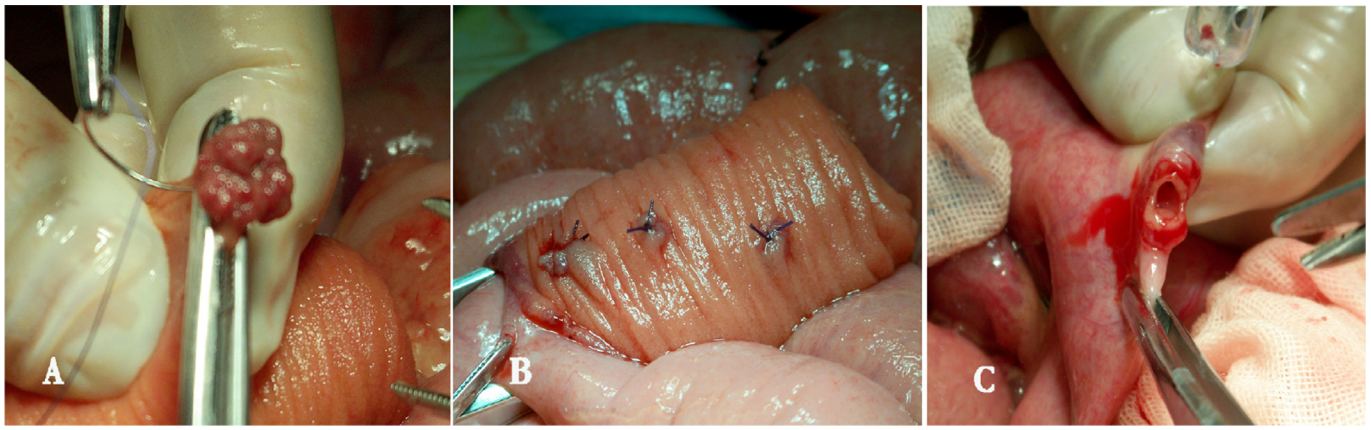

Fig. 3. The different measures for dealing with lesions in GI tract. A: Cross-suturing ligation at the bottom of lesion for interruption of blood supply. B: A technique of bowel intussusception was used to access multiple venous malformations. C: lesion was resected.

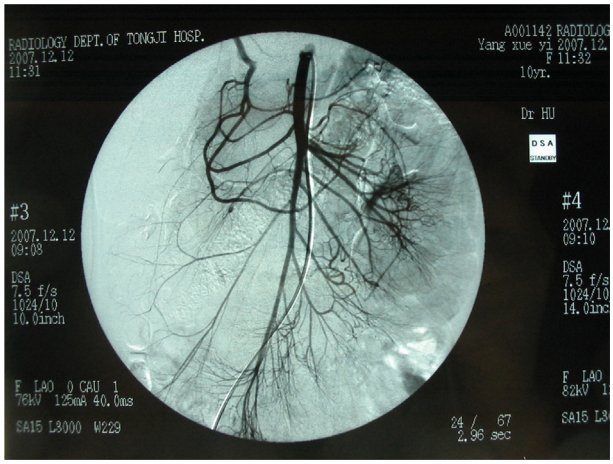

Fig. 4. DSA.

(Fig. 1A–C); A small amount of active bleeding at the right lower quadrant was displayed by SPECT examination. Small intestine dual-phase scan and reconstruction of CT indicted a number of nodules in right lobe of liver and in small bowel wall. DSA also presented a number of abnormal small staining points in liver, intestinal and stomach (Fig. 3)

A laparotomy more than 10 h was performed. VM lesions were widely distributed over stomach, small intestine and colon. Most of them located on the intestinal mucosa which appeared as the strawberry-shaped protrusions. Some of lesions penetrated bowel wall into serosa side (Fig. 2B). The VM lesions resulted in two intussusceptions in ileum (Fig. 2C).

The stomach cavity was opened and gastric lesions were resected. From beginning of the treitz ligament, the trans-wall lesions were examined segment by segment. A segment of 40 cm terminal ileum was resected because of a number of lesions. A total of 81 lesions were dealt with. Among them, 52 were resected and 29 were ligated. One gastric anastomosis and eleven intestinal anastomoses were performed. Followed-up 3 months after this surgery, patient had occasionally black stool, but hemoglobin still less then 100 g/L.

Four months after the first surgery, the second surgery was performed.5 lesions in duodenum and 13 lesions in colon were removed. One-year follow-up was done after the secondary surgery. A satisfactory effect had been achieved, and hemoglobin was maintained over 120 g/L without any iron supplementary or blood transfusion.

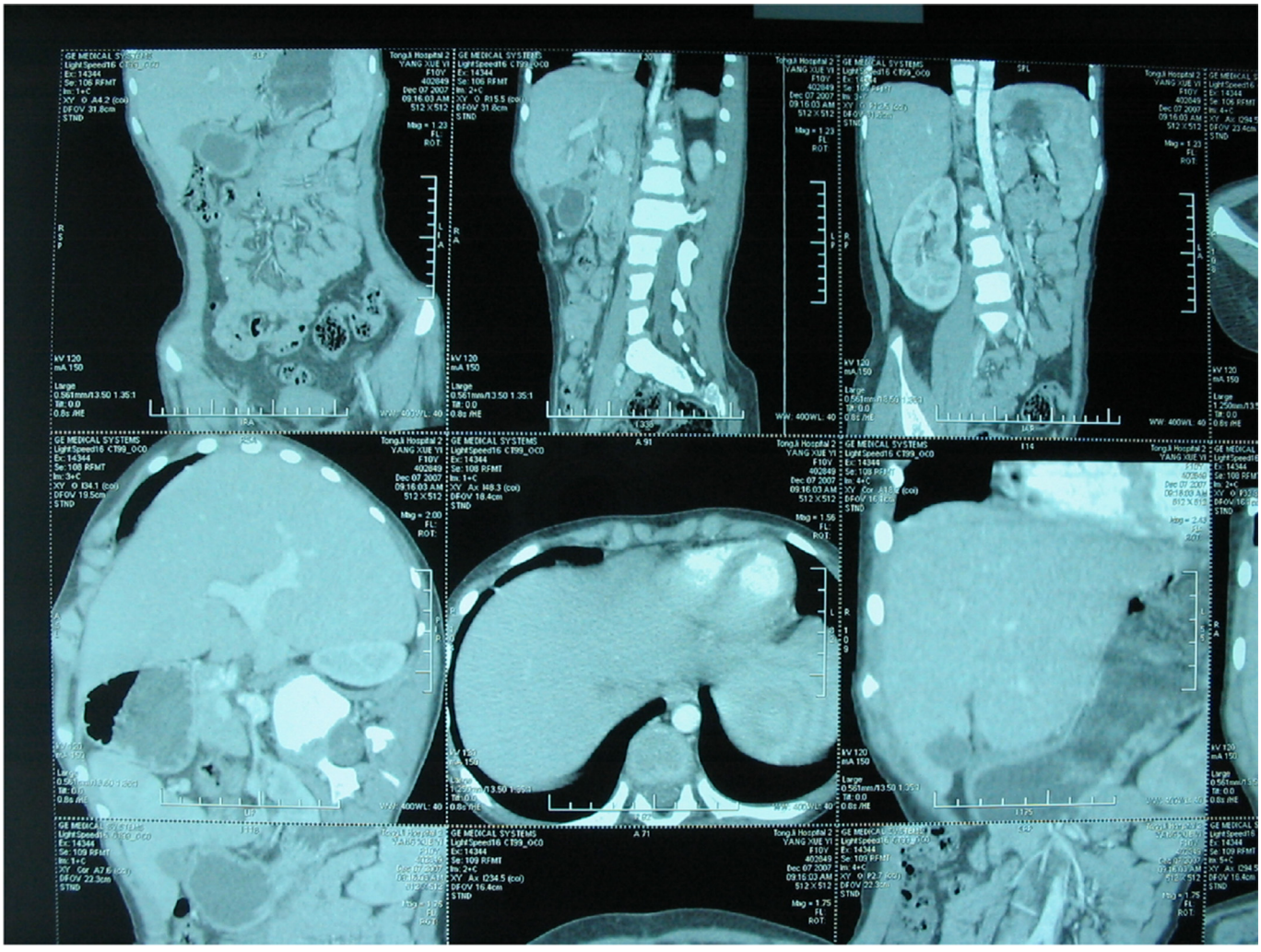

Fig. 5. CT scan.

3. Discussion

Blue rubber bleb nevus syndrome (BRBNS) is a rare vascular anomaly syndrome consisting of multifocal venous malformations (VM) of the skin, soft tissues, and gastrointestinal (GI) tract [1–6]. Due to its low incidence and the hemangioma-like appearance, clinicians often made an incorrect diagnosis. For example, the VM in skin were easily diagnosed as hemangioma. Our two patients had undergone superficial hemangioma resection when they were at an early age.

Although VM involve in many tissues and organs, GI bleeding is the main clinical problem. GI bleeding was manifested as intermittent melena at the initial stage, and then became gradually severe and required repeating blood transfusion. For VM in GI tract, endoscopy examination is the most valuable diagnostic methods (Fig. 1A–C). However, endoscopic operator could make incorrect diagnosis and delayed treatment because of lacking experiences [4,5].

DSA and SPECT examinations were also performed in these two patients. However, both DSA and SPECT examinations could not make an accurate diagnosis for VM, only for tracing GI bleeding sites (Fig. 4). Dual-phase CT scan of small intestine could only find some small nodules (Fig. 5). In generally, DSA, SPECT and CT are more sensitive to a large amount of arterial hemorrhage, but we found that the diameter of VM lesions was usually less than 1 cm without active bleeding. Therefore, we believed that endoscopy should be more valuable for diagnosis.

There was no consensus on treatment of the VM in GI tract. Because chronic GI bleeding persisted throughout all life, the treatment for lesions in GI tract were most important. A variety of pharmacological therapeutic strategies, including antiangiogenic agents and interferon, have been proposed for GI bleeding of BRBNS. However, these agents have not been demonstrated to be reliable and effective for reducing and controlling GI bleeding [4–7].

Recently, Fishman and other researchers believed BRNBS lesions are congenital vascular malformation rather than proliferation tumor. Based on this theory, Fishman treated 10 cases BRNBS patients with surgery, an average of 137 lesions (4–557) were resected in each patient, a small amount of bleeding recurrence without severe anemia in one patient, further surgical intervention in two patients [6]. These two cases also demonstrated that surgical eradication for VM lesions was important and effective for preventing GI bleeding.

From our experience, we believe that serious chronic bleeding from GI venous malformations of BRBNS can be successfully controlled by aggressive resection of the VM, regardless of their number or location. Pharmacologic therapy is unlikely to be of benefit. This aggressive operative approach can help avoid the ravages of chronic anemia, including fatigue, iron overload, and the potential risks of repetitive blood transfusions including hepatitis and allosensitization.

Surgical intervention is best planned when a transfusion requirement becomes apparent. Once an operative approach is chosen, complete visualization of the entire gastrointestinal mucosa from the mouth to the anus must be performed. Complete eradication may avoid the need for repeated operations. Incomplete removal of any given lesion may allow for reexpansion and recurrent bleeding. Although extremely tedious, standard surgical principles and techniques combined with careful anesthetic management allow for a successful outcome with minimal morbidity.

Patients with an overwhelming number of lesions may require staged operative procedures. Nevertheless, this approach dramatically improves the health and well-being of patients with blue rubber bleb nevus syndrome (BRBNS).

Ethical approval

Tongji Hospital Ethical Committee.

Sources of funding

This study was funded by National Natural Science Foundation of China. (PI: Zhi Li; No: 81400579 and PI: Yongjun Chen; No:81372239).

Author contribution

Xiaoyi Sun designed this study.
Zhi Li collected data and wrote this manuscript.

Conflicts of interest

No conflicts of interest.

Guarantor

Zhi Li,Xiaoyi Sun.

Acknowledgment

This study was funded by National Natural Science Foundation of China. (PI: Zhi Li; No: 81400579 and PI: Yongjun Chen; No: 81372239 and 81572783).

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