

## Case Report

# Unusual Cause and Association of Gastrointestinal Bleed in a Young Boy

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**ABSTRACT** A 4-year old child, previously operated case of tetralogy of Fallot present with recurrent episodes of massive lower gastrointestinal bleed of one year duration. Endoscopic evaluation revealed multiple bluish vascular lesions in the duodenum and proximal jejunum and a single rectal polyp. Histology of the lesion was suggestive of venous malformation consistent with blue rubber bleb nevus syndrome (BRBNS). Child underwent endoscopic snaring and surgical resection with end to end anastomosis. At six months' follow-up child was asymptomatic without any bleed episodes.

**KEYWORDS:** Blue rubber bleb nevus, child, gastrointestinal bleed

## INTRODUCTION

Gastrointestinal (GI) bleed is one of the most common GI emergencies in children.<sup>[1]</sup> At times, identification of the source of bleed is challenging. Apart from common causes of rectal bleed such as polyp, rectal ulcer and Meckel's diverticulum, less common sources in the small intestine like vascular malformation would pose difficulty in confirmation and planning an appropriate treatment. We here present a rare case of multifocal vascular malformation with an unusual association of tetralogy of Fallot, who was treated vascular malformation by combined endoscopic and surgical approach.

## CASE REPORT

A 4-year-old boy presented with 4 episodes of massive melena in the past one year requiring at least 2 units of packed red cell transfusions each time. The child was cyanotic since early infancy, diagnosed as a case of tetralogy of Fallot and surgically corrected at 4 years of age. He also complained of a swelling in the left upper thigh which was present since birth and was progressively increasing. There was no history of hematemesis or bleeding from any other site. At a presentation to the emergency room, the child was in compensated shock with pallor, hemoglobin of 6.3 g% for which he received packed red blood cells at 20 ml/kg body weight. His anthropometry (weight: 14.5 kg and height

103 cm) is between 0 and -2 standard deviation scores for age. A sternotomy scar of previous cardiac surgery was noted. A 4 cm × 5 cm asymptomatic elevated skin lesion in the upper left thigh was present, which was later confirmed to be venous malformation. Systemic examination was normal. Esophagogastroduodenoscopy revealed multiple bluish pedunculated polypoidal lesions in the duodenum and proximal jejunum measuring up to 10 mm in size [Figure 1]. Ileo-colonoscopy examination did not reveal any vascular lesion except for a similar polypoidal lesion in the rectum. The child underwent endoscopic snaring of the duodenal and proximally jejunal vascular lesions and the rectal polyp. Histology of all the polyps was similar. There were multiple dilated thin wall vascular channels expanding the lamina propria extending across the glands and to the submucosa, which were included in the biopsies. These vascular channels were confirmed by CD31 on immunostaining [Figure 2]. The child was labeled as a case of Blue Rubber Bleb Nevus Syndrome (BRBNS) based on the clinical and endoscopic features, and the typical morphological finding of the mucosal polypoidal lesion. A month later, he was once again admitted with massive lower GI bleed and taken for emergency

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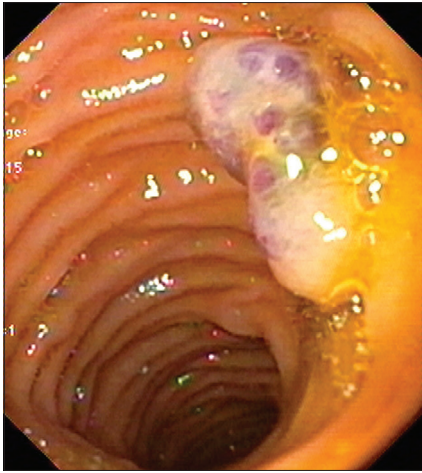
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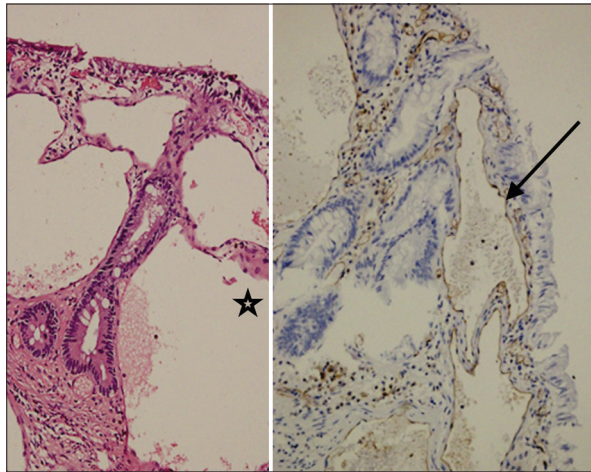


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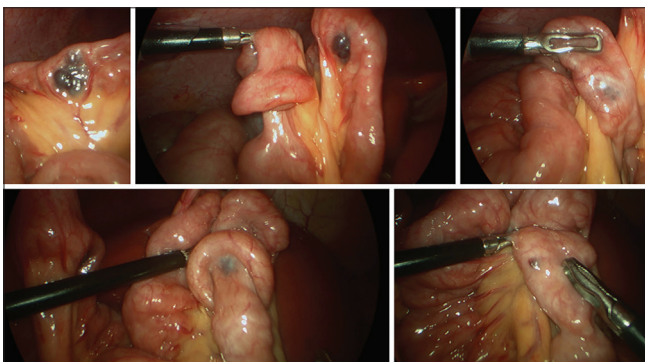
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**Figure 1:** A bluish pedunculated polypoidal lesions in the duodenum suggestive of venous malformation



**Figure 2:** Multiple dilated thin wall vascular channels expanding the lamina propria (asterisk). CD31 positivity of the endothelial lining suggesting vascular origin (arrow)



**Figure 3:** Transmural bluish colored vascular lesions demonstrated in the small intestine during laparoscopy

intraoperative enteroscopy. During laparoscopy, about 20 transmural bluish colored vascular lesions were noted, predominantly in the jejunum [Figure 3]. Of these twenty lesions, 15 were wedge resected, and the larger transmural lesions were removed by complete

resection followed by end-to-end anastomosis of bowel. A total of 10 cm of bowel was removed. There were no residual lesions. Recovery was uneventful. Histology of the resected intestinal segments showed the similar morphology of the polypoidal mucosal lesion. At six months, postsurgery child was asymptomatic without any bleeding episodes.

## DISCUSSION

BRBNS is a rare venous type of vascular malformation involving the skin, GI tract and musculoskeletal system followed by other organs like the brain.<sup>[2]</sup> Lesions are categorized according to the criteria of International Society for the Study of Vascular Anomalies, which involves both gross and histological features. Exact etiology of BRBNS is unclear. However, defect in the morphogenesis of the venous development has been postulated.<sup>[3]</sup> The majority of the cases are sporadic, but there are reports of autosomal transmission.<sup>[4]</sup> GI lesions are clinically significant compared to other sites due to the associated risk of chronic blood loss and anemia or at times brisk massive life-threatening bleed.<sup>[5]</sup> Other less common presentations are intussusception, luminal obstruction in large lesions and intestinal infarction of the bowel. Histologically, they appear as dilated venous channels and rarely phlebolith formation due to sluggish venous blood flow. Exact characterization of the lesion is important as this has to be differentiated from other vascular neoplasms like capillary hemangioma in infants and younger children. There is no consensus on the exact management, but it depends on the site of lesion, associated complications, and expertise of the treating physician. Large GI lesions are excised by laser photocoagulation or endoscopic polypectomy. Transmural and lesions spanning over a significant length of the intestine would require surgical resection. Recently, an anecdotal report claims response to sirolimus at dose of 0.05–0.1 mg/kg/day.<sup>[6]</sup> The association of this condition with congenital heart disease is very rare with only a single case report so far.<sup>[7]</sup> To the best of our knowledge, this is the first report of a case documenting BRBNS associated with tetralogy of Fallot.

## CONCLUSION

BRBNS is a multivisceral venous malformation. GI lesions are one of the common afflictions with significant morbidity. A combined approach as highlighted above would improve the outcome with the minimal small intestine loss.

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## Conflicts of interest

There are no conflicts of interest.

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