

Case Report

Diffuse hemangiolympangioma of small bowel-rare cause of obscure gastrointestinal bleeding

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ABSTRACT

Hemangiolympangioma (HL) is a rare benign tumor, commonly present in pediatric age group that involves the head and neck. Small bowel HL has been reported as solitary or segmental involvement. Here we report a case of diffuse small bowel HL, presented as chronic anemia, fatigability and obscure gastrointestinal (GI) bleeding. A 42-year male presented with 15 years history of bleeding per rectum, anemia, and fatigability, received multiple blood transfusions. The exact cause could not be identified. His complete hemogram parameters were low requiring further imaging studies. Imaging and intraoperative enteroscopy showed multiple nodular lesions in mesentery as well as in the mucosa of the entire small bowel. Bleeding points were noted in a proximal jejunal lesion which was resected. Histopathological study confirmed as benign HL. HL are rare tumors of gastrointestinal tract. To the best of our knowledge diffuse HL of the small bowel have not been reported in the medical literature. With this case scenario, one should consider these tumors in the differential diagnosis of obscure lower GI bleed and chronic anemia, as well as explore definitive management options for these benign symptomatic tumors.

Keywords: HL, Small bowel, GI bleeding

INTRODUCTION

Hemangiolympangioma (HL) is a rare benign tumor, commonly present in pediatric age group that involve head and neck. Small bowel HL has been reported as solitary or segmental involvement. Here we report a case of diffuse small bowel HL, presented as anemia, fatigability and obscure GI bleeding.

HL is a benign tumor of vascular malformation. The proposed theory for the formation is obstruction of venolymphatic communication, dysembryoplastic lymphatic and vascular tissue.¹ Intramural lymphatic obstruction, endothelial permeability damage, swelling, and congenital absence of lymph have been proposed as

other possible causes for the development of small bowel lymphangiomas.²

HLs are nodular lesion with cystic walls lying inside dilated lymphatic spaces and attenuated by endothelial cells. The cyst fluid can be chylous, hemorrhagic, or serous contained within a collagen stroma and abundant eosinophilic granules.³

Most of the intestinal HLs are incidentally discovered, during an endoscopy or a radiological study.⁴⁻⁶

Herein, we report a case of diffuse nodular HLs of the small bowel presented with obscure lower gastrointestinal bleeding.

CASE REPORT

A 42-year-old male presented with easy fatigability and occasional bleeding per rectum. He was evaluated several times in various hospitals, received multiple blood transfusions, but exact cause couldn't be identified. He underwent hemorrhoidectomy 10 years ago. In 2019 he was admitted with massive bleeding per rectum, severe anemia and congestive cardiac failure. On examination he was pale, no clubbing, and no generalized lymphadenopathy. Per abdomen examination revealed exaggerated bowel sounds otherwise unremarkable. Per rectal examination showed bleeding. Upper GI scopy showed pale gastric mucosa. Colonoscopy showed a polyp in ascending colon which was not actively bleeding and terminal ileitis. CECT abdomen revealed contrast extravasation near ileum. RBC tagged Technetium 99m isotope scan showed blood pooling near ileum. On laparotomy there was multiple submucosal nodular lesions in the small bowel and Meckel's diverticulum. Intraoperative enteroscopy revealed multiple mucosal nodular lesions in small bowel. Since there was no active bleeding on enteroscopy and preoperative investigations (CECT+ isotope scan) showed bleeding from the region of ileum, Meckel's diverticulectomy and incisional biopsy from mesenteric nodular lesion reported as fibromatous tissue. Postoperatively he developed hemorrhagic ascites with chyle and bleeding per rectum. Repeat CECT abdomen showed prominent mucosal folds in jejunum and multiple small nodular lesion seen in jejunal lumen and mesentery with gross ascites (Figure 1 and 2).

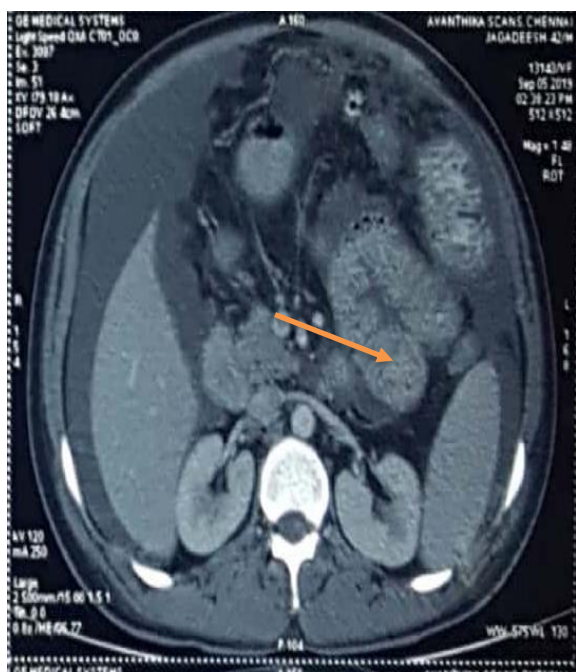


Figure 1: CECT abdomen with oral contrast (Axial section) showed prominent mucosal folds in jejunum and multiple small nodular lesions seen in jejunum (arrow) and gross ascites.



Figure 2: CECT coronal section revealed multiple mesenteric nodular lesion (arrow) along with mucosal nodules in small bowel.

Based on previous intraoperative findings and present CT report, experts were consulted and collectively diagnosis of HL was made. It was decided to do re-laparotomy and intraoperative enteroscopy. On relaparotomy there was multiple nodular lymphangiomatous lesion present in mesenteric border of small bowel. Three liters of hemorrhagic ascites with chyle drained. Bleeding points noted in proximal jejunum intra-luminally on enteroscopy. The Whole mesentery and mesenteric side of the small bowel were studded with nodular yellow HL tous lesion (Figure 3). Proximal 50 cm of jejunum was resected and end-end anastomosis was done (Figure 4).

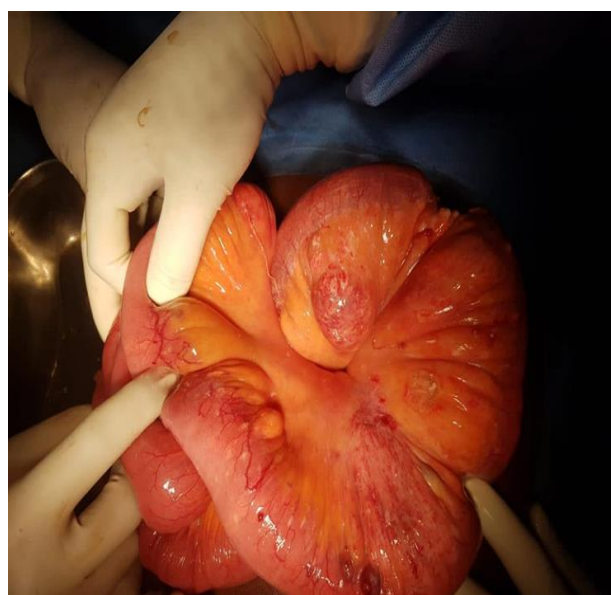


Figure 3: Intraoperative image of multiple mesenteric nodules.



Figure 4: Resected jejunal specimen of multiple sub-mucosal nodules suggestive of HL.

Feeding vessel of rest of HLs were ligated without compromising viability of small bowel. Final histopathology report showed sub serosal and focally in submucosal multiple closely packed thin-walled dilated vascular channels of variable size, lined by flat endothelial cells. Lumen shows serous fluid and hemorrhage with intervening areas shows congested vessels, adipose tissue, fibromuscular tissue and lymphoid aggregates as benign vascular lesion suggestive of HL (Figure 5). No evidence of malignancy found. Postoperatively he developed chylous ascites which was managed conservatively with medium chain triglycerides (coconut oil). Chylous ascites settled. He remained asymptomatic till now.

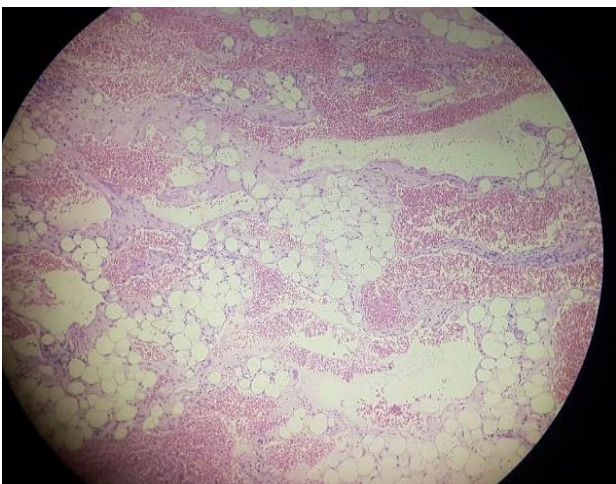


Figure 5: Biopsy of resected jejunum shows serosal and sub-mucosal thin-walled dilated vascular channels of variable size, lined by flat endothelial cells. Lumen shows serous fluid and hemorrhage with intervening areas shows congested vessels, adipose tissue, fibromuscular tissue and lymphoid aggregates as benign vascular lesion suggestive of HL.

DISCUSSION

HLs is a very rare benign tumor with incidence varies from 1.2 to 2.8 per 1000 newborns, and no gender predilection.¹ HL can be of primary which is congenital malformation of vascular and lymphatic system, or secondary to injury of lymphatic vessels following trauma or operation, which induces inadequate lymph fluid drainage. On gross morphology it can be classified as cystic or cavernous lesion. Microscopically, the tumor consists of multiple closely packed thin-walled dilated vascular channels lined by flat endothelial cells and lymphatics. In small bowel HLs, submucosal and subserosal vascular and lymphatic network is affected, with many dilated, thin-walled and irregular blood and lymph fluid filled spaces, mainly located within mucosa, submucosa as well as in mesenteric vascular channels.

HLs of small bowel may present as intermittent moderate to massive painless lower GI bleeding which complicate as anemia and easy fatigability. Clinical examination are usually remains elusive. Upper GI scopy and colonoscopy should be done to rule out accessible causes of GI bleeding. CT or MRI are useful in identifying the source of bleed and possible site of the lesion. However, it is very difficult to diagnose preoperatively.

Complete resection is the definitive treatment for solitary or segmental involvement of bowel. In case of diffuse involvement of small intestine, actively bleeding segment of intestine can be resected and rest of the lesion can be managed by ligating the feeding vessel without compromising the vascularity of bowel. Other non-surgical options include sclerotherapy, electrocautery, radium implantation, cryosurgery and laser therapy can be tried to control bleeding, but these may only result in temporary clinical outcomes.^{7,8} Angiographic embolization can also be applied in case of solitary lesion with feeding vessel. Medical management like steroids, hormones may be tried for regression of tumor, but often not effective.

CONCLUSION

HL s are rare tumors of gastrointestinal tract. To the best of our knowledge diffuse HLs of small bowel has not been reported in the medical literature. With this case scenario, one should consider these tumors in the differential diagnosis of obscure lower GI bleed and chronic anemia, as well as explore definitive management options for these benign symptomatic tumors.

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