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International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Small bowel lymphangioma causing ileo-ileal intussusception in adults



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ARTICLE INFO

Article history:

Received 23 October 2017

Received in revised form

14 November 2017

Accepted 16 November 2017

Available online 21 November 2017

Keywords:

Small bowel lymphangioma

Ileo-ileal intussusception

Abdominal pain

Cystic congenital malformation

Lymphatic system

ABSTRACT

INTRODUCTION: Lymphangioma is a rare benign tumor found in gastrointestinal tract. Most lymphangiomas can occur at any age and but mostly are found in children and infants. They are mainly due to malformation of the lymphatic system. They occur mainly in the head, neck and oral cavity, but less commonly develop in the abdominal cavity. Colonoscopy and endoscopic ultrasonography are frequently used to diagnose lymphangiomas of the small bowel. Ileo-ileal intussusception due to small bowel lymphangiomas has been rarely reported.

PRESENTATION OF CASE: We report a case of 24 year old female who presented to the hospital with sudden onset of right sided upper and lower abdominal pain with nausea and vomiting, elevated WBC count of 15.1 μ L. After careful examination and CT scan of Abdomen patient was found to have an ileo-ileal intussusception secondary to small bowel lymphangioma. In a rare clinical presented case, we performed a diagnostic laparoscopy (Fig. 1), exploratory laparotomy, small bowel resection, and stable primary anastomosis. Post-operative patient did well and was discharged on 4th post-operative day without complications.

DISCUSSION: Lymphangiomas are rare benign tumors which have soft tissue consistency and often congenital malformation of the lymphatic system. Ileo-ileal intussusception in an adults can be a life threatening condition which requires prompt diagnosis and urgent intervention due to the risk of developing ischemic bowel. Prompt diagnosis and urgent intervention can lead to favourable

CONCLUSION: We present this rare case of ileo-ileal intussusception secondary to small bowel lymphangioma with literature review.

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1. Introduction

Lymphangioma is a rare benign tumor which are found in gastrointestinal tract. Once they occur they cause severe right sided abdominal pain. Most lymphangiomas can occur at any age and but mostly are found in children and infants. They are mainly due to malformations of the lymphatic system. They occur mainly in the head, neck and oral cavity, but less commonly develop in the abdominal cavity. This case report has been reported in line with SCARE criteria [1].

2. Case presentation

A 24 year old female presented to the hospital with sudden onset of right sided upper and lower abdominal pain with some nausea

but no vomiting. Patient had no fever with stable blood pressure and heart rate. Abdominal examination was positive for Right sided abdominal tenderness. Laboratory tests showed an elevated WBC count of 15.1 μ L with no other significant lab changes. CT of the abdomen/pelvis was obtained which showed internal hernia at the mid pelvis, involving loop of ileum (Fig. 2). There was presence of mesenteric fat within a loop of bowel which suggested intussusception with possibility of small bowel mass. Patient was taken to operating room for diagnostic laparoscopy. Intra-operatively, an ischemic loop of bowel was identified secondary to internal herniation or intussusception, laparoscopy was converted to exploratory laparotomy, confirming the presence of ischemic bowel secondary to intussusception with questionable mass/cystic structure. Small bowel resection and primary anastomosis was completed. The size of the resected mass was measured to be 28 cm \times 15 cm (Fig. 1). Excised specimen had a gross appearance of the small bowel showing invagination of an intestinal segment into the next part of the small bowel hence it was consistent with the ileo-ileal intussusception pathology was positive for cystic lymphangioma confirming the diagnosis of small bowel lymphangioma (Fig. 3). Study of the

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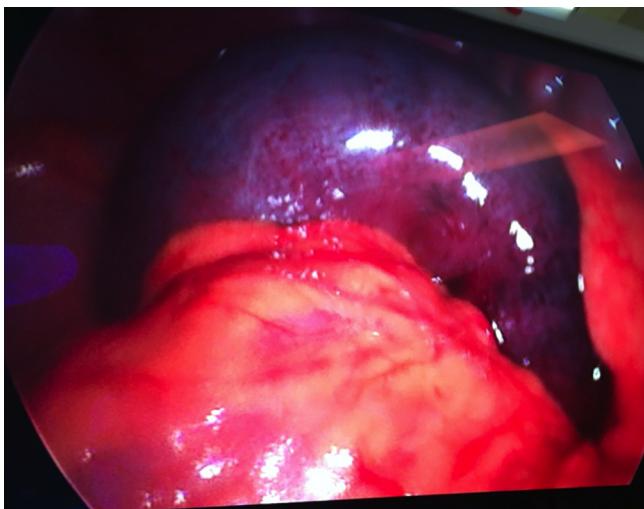


Fig. 1. Laparoscopic Findings: Lymph vessels are dilated as seen in the ileal region of the small bowel and also had very rubber type of texture feeling when touched with forceps.

histology slide of the tumor which mainly showed area of cystic lymphangioma, irregular cysts with walls composed of smooth muscle cells and fibrocytes, and the inner surface was covered with flat endothelial epithelium, dilated lymphatic lacunae, abundant lymphatic lymphocytes (Fig. 4). Post-operative patient did well and was uneventful and she was discharged on 4th post-operative day without any post-operative complications. Patient came for a 2 weeks follow up and was doing well tolerating diet, having normal bowel movements with mild incisional site pain and incisional wound healing well.

3. Discussion

Lymphangiomas are rare benign, soft-tissue, cystic congenital malformation of the lymphatic system, typically occurring in children [9]. They usually present as microscopic or macroscopic vesicles and channels filled with clear to serosanguinous fluid. They most typically occur in infants and children. Most occur in the head and neck, only 10% occur in internal organs [4]. Fewer than 1% occur in the mesentery, omentum, and retroperitoneum [2]. Abdominal lymphangiomas are most commonly found in the mesentery of small bowel (80%), and have been suggested to result from an embryological failure of the lymphatics when primary lymphatic sacs fail to join the lymphatic system [2]. Other possi-



Fig. 3. Excised Specimen: This gross image shows thinned out wall and dark brown mucosa cover with clotted blood. A broad based polypoid lesion with cyst measuring 1.7 cm in greatest dimension. Also gangrenous portion of intussuscepted small bowel.

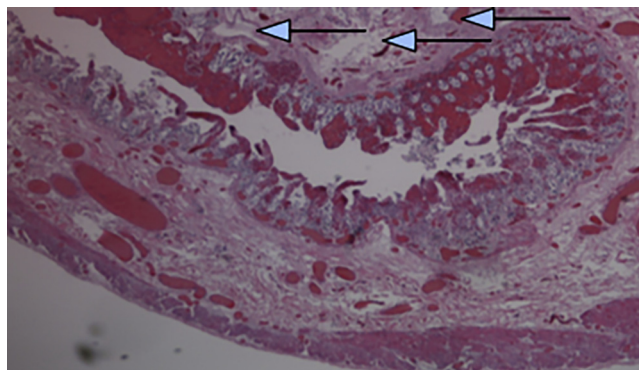


Fig. 4. Histopathological findings: arrows are pointing to the area of cystic lymphangioma, irregular cysts with walls composed of smooth muscle cells and fibrocytes, and the inner surface was covered with flat endothelial epithelium, dilated lymphatic lacunae, abundant lymphatic lymphocyte, or lymphoid tissues.

ble theories include acquired inflammation of lymphatic channels leading to obstruction and subsequent cystic lymphangioma [8]. This has been supported by an in vivo study in which corneal lymphangiogenesis was induced by thermal cauterization, in a



Fig. 2. CT of Abdomen: abnormal loop of small bowel in the pelvis. The presence of mesenteric fat within what appears to be a loop of bowel suggests intussusception.

mouse cornea. Thermal cauterization simulated lymphangiogenesis in inflammatory conditions, such as keratitis, chemical burns and graft rejection. Additionally, in this study it was found that matrix metalloproteinase (MMP) is a key regulator in the remodeling process of lymphangiogenesis by the inhibition of lymphatic endothelial cells (LEC) sprouting achieved by using a synthetic MMP inhibitor [3]. Histopathologically, lymphangiomas can be divided into 3 groups: cystic, simple and cavernous lymphangiomas. Cystic lymphangiomas on histology appeared as irregular cysts with walls composed of smooth muscle cells and fibrocytes, and the inner surface was covered with flat endothelial epithelium, dilated lymphatic lacunae, abundant lymphatic lymphocyte, or lymphoid tissues [10]. Also, the human endothelial marker podoplanin, which is recognized by the monoclonal antibody D2-40 has been identified as a useful marker for lymphatic neoplasms [8].

Clinical presentation of abdominal lymphangioma is diverse, however typically patients are asymptomatic or have nonspecific symptoms [2]. Clinical signs and symptoms include intermittent, cramping abdominal pain, diarrhea, fullness, abdominal mass. GI bleed occurs in cases in which abdominal lymphangioma occurs within the bowel. Melena occurs when lymphangioma occurs in the small bowel or above, whereas bright red blood will occur with lymphangiomas occurring within the colon and rectum. Protein-losing enteropathy is specific for lymphangioma occurring within the bowel [6]. Volvulus typically occurs when lymphangiomas is present on the mesentery of the bowel, while intussusception occurs when lymphangioma is present on mesentery or on or within the bowel itself. In the pediatric population, males seem to be more commonly affected by lymphangiomas. However, in the adult population, females seem to be more commonly affected. It was proposed in the study “Intra-abdominal and retroperitoneal lymphangiomas in pediatric and adult patients” that in the adult population endogenous estrogens might be involved in causing enlargement or growth of lymphangiomas, explaining the prevalence of intra-abdominal lymphangiomas in adult females. In “Gastrointestinal tract lymphangiomas: findings at CT and endoscopic imaging with histopathologic correlation,” 4/6 patients were female. However, in both studies, sample sizes were small [5,10].

In a retrospective study from 2007, characteristic imaging findings for gastrointestinal tract lymphangiomas were described, with CT images for 6 patients and endoscopic patients for 4 patients. On CT, they showed a characteristic appearance with a well-demarcated, thin-walled oval mass of low attenuation beneath the submucous membrane. These masses showed a stratifying effect because the cyst is homogenous, watery, and non-enhancing and lies between enhanced mucous membrane and serous membrane. The diameter of the cystic lesions ranged from 0.8 cm to 3.5 cm. Two patients had associated intussusception, which was evident on CT as thickened intestine wall and soft tissue and fat attenuation within the intestine lumen producing concentric layering effect. Another patient with lymphangioma occurring in bowel mesentery, with associated intestinal volvulus had whirlpool sign on CT, a finding consistent with volvulus. On endoscopy of lymphangiomas in the small intestine, endoscopic photographs showed submucosal mass, cushion sign, and alteration in shape as patient changed positions. These lesions were further described as being covered with healthy and typical-appearing mucosa with distended mucosal vessels, blue-tinged, soft, and easily deformed by pressure. In this research study, it is proposed that CT and endoscopy are valuable tools to diagnose and manage gastrointestinal lymphangiomas [10].

To our knowledge and with literature review there has been one more case reported of ileo-ileal intussusception has occurred due to lymphangioma in an adult [7]. The definitive treatment for gastrointestinal cystic lymphangioma is resection, with resection of the involved segment of bowel the most preferred procedure.

However, resection of the lymphangioma can be performed endoscopically or surgically. Typically, endoscopic resection is performed for lymphangiomas 2 cm or less in the maximal diameter [8]. In certain cases, cysts cannot be completely removed without injuring contiguous viscera, in which case there are two alternative treatments. Firstly, marsupialization and sclerosis of the cyst lining may be performed, due to the fact that partial excisions alone are associated with a high rate of recurrence. If it is not possible to completely remove the cyst, sclerosing agents may be used to shrink the cyst [10]. Such therapies include bleomycin and sodium tetradecyl sulfate [1].

4. Conclusion

Ileo-ileal intussusception in an adult can be a life threatening condition as it risks bowel ischemia. Prompt diagnosis and urgent intervention can lead to favourable outcome. To our knowledge this is a rare case report of ileo-ileal intussusception secondary to small bowel lymphangioma in an adult.

Conflicts of interest

No conflicts.

Funding

No source of funding.

Ethical approval

Ethical approval is not required by our institution. This was a case report and permission and consent has been taken from the patient.

Consent

We have obtained a written informed consent from the patient for publication of this case report and also accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Authors contribution

Khuram Khan – Abstract, Figure collections, writing, format.
Lauren Kleess – writing, others.
Ram Ganga – others, writing, format.
Hector DePaz – review, final writing, editing.
Robert Santopietro – other, editing.

Guarantor

Khuram Khan.

References

- [1] R.A. Agha, A.J. Fowler, A. Saetta, I. Barai, S. Rajmohan, D.P. Orgill, for the SCARE Group, The SCARE statement: consensus-based surgical case report guidelines, *Int. J. Surg.* (2016).
- [2] K.M. AlGhamdi, T.F. Mubki, Treatment of lymphangioma circumscriptum with sclerotherapy: an ignored effective remedy, *J. Cosmet. Derm.* 10 (2011) 156–158.
- [3] B. Detry, F. Bruyere, C. Erpicum, Digging deeper into lymphatic vessel formation in vitro and in vivo, *BMC Cell Biol.* 12 (2011) 29.
- [4] D.L. Grasso, G. Pelizzo, E. Zocconi, J.G. Schleeff, Lymphangiomas of the head and neck in children, *Acta Otorhinolaryngol. Ital.* 1 (2008) 7–20, PMID: PMC2640069.
- [5] B.K. Goh, Y.M. Tan, H.S. Ong, Intra-abdominal and retroperitoneal lymphangiomas in pediatric and adult patients, *World J. Surg.* 29 (2007) 837–840.

- [6] K.L. Hugeot, P.P. Metzger, D.M. Menke, Colorectal lymphangioma, *Am. Surg.* 73 (2007) 414–416.
- [7] A. Kohga, A. Kawabe, Y. Hasegawa, et al., Ileo-ileal intussusception caused by lymphangioma of the small bowel treated by single-incision laparoscopic-assisted ileal resection, *World J. Gastroenterol.* 23 (1) (2017) 167–172, <http://dx.doi.org/10.3748/wjg.v23.i1.167>.
- [8] A.Y. Mehmet, A. Sezer, E. Yeldan, Lymphangiomas of the colon mimicking acute appendicitis, *Indian J. Cancer* 48 (2011) 139–140.
- [9] R. Mendez-Gallart, A. Bautista, E. Estevez, P. Rodriguez-Barca, Abdominal cystic lymphangiomas in pediatrics: surgical approach and outcomes, *Acta Chir. Belg.* 111 (2011) 374–377;
- [10] S. Vallance, Giant cystic lymphangiomas, *ANZ J. Surg.* 80 (2010) 292–299.
- [10] H. Zhu, Z.Y. Wu, X.Z. Lin, Gastrointestinal tract lymphangiomas: findings at CT and endoscopic imaging with histopathologic correlation, *Abdom. Imaging* 33 (2007) 662–668.

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