

Case Report

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Giant mesenteric cystic lymphangioma: a rare cause of intra-abdominal catastrophe

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ABSTRACT

Lymphangiomas are uncommon benign tumours and occur mainly in children with the most common sites being head and neck followed by the axilla and mediastinum. Intra-abdominal lymphangiomas are rare. Clinical presentation is diverse ranging from incidentally discovered abdominal mass to symptoms of acute abdomen. Life threatening complications are more likely to develop in children. We here present a rare case of giant mesenteric cystic lymphangioma causing intra-abdominal catastrophe in an adult.

Keywords: Cystic lymphangioma, Gut gangrene, Mesentery

INTRODUCTION

Lymphangiomas are uncommon benign tumours and occur mainly in children with 90% of these manifesting in children younger than 3 years.^{1,2} The most common sites being head and neck followed by the axilla and mediastinum. Intra-abdominal lymphangiomas are rare.³ Clinical presentation is diverse ranging from incidentally discovered abdominal mass to symptoms of acute abdomen. Children are more likely to develop life threatening complications.^{4,5} We here present a rare case of giant mesenteric cystic lymphangioma causing intra-abdominal catastrophe in an adult.

CASE REPORT

A 34-year old gentleman presented to our casualty with complaints of sudden onset pain abdomen, distension of abdomen and obstipation for 1 day. There was no history of fever or vomiting or any history of previous surgery. Patient gave history of colicky pain in the umbilical

region on and off for last 6 months. On examination, patient was dehydrated, pulse – 112/ min, B.P. – 134/86 mm Hg. Abdomen was tense, distended and tenderness and guarding was present all over, entire abdomen was tympanic except for dullness in bilateral lumbar and iliac regions. Bowel sound was absent. Peritoneal tapping over area of dullness revealed foul smelling haemorrhagic fluid. A provisional diagnosis of gut gangrene was made and patient was posted for emergency laparotomy after resuscitation with i.v. fluids and i.v. antibiotics.

Intraoperatively, there was around 0.5 litre of foul smelling haemorrhagic fluid, the small bowel of length around 100 cm, 6 feet distal to the duodenal-jejunal (DJ) flexure up to the terminal ileum was twisted 360° clockwise and found to be gangrenous. There was a yellowish- white multi-loculated lesion (Figure 1) of size 20×12×5 cm in the mesentery in close proximity and adjoining the small bowel around 8 feet distal to the DJ flexure, adhered to the parietal peritoneum near the inferior border of right lobe of liver. The lesion was

excised intoto with gangrenous small bowel, appendix and caecum and ileo-ascending colon anastomosis was fashioned. Histopathological examination revealed multiple dilated lymphatic spaces in the cyst wall lined by a single layer of cuboidal to flat epithelium with stroma consisting of loose fibro-vascular tissue and smooth muscle. Features suggestive of cystic lymphangioma.



Figure 1: Resected specimen showing the lesion in the mesentery along with the gangrenous segment of small bowel.

DISCUSSION

Mesenteric cystic lymphangiomas are rare benign tumours of the lymphatic system with a reported incidence of less than 1 per 100000 hospital admissions.^{6,7}

The exact etiology remains unclear. A well-established theory suggests that lymphangiomas arise from sequestrations of lymphatic tissue during embryologic development.² However it is suggested that abdominal trauma, lymphatic obstruction, inflammatory process, surgery or radiation therapy may lead to secondary formation of such a tumour.¹

Mesenteric lymphangiomas are either incidentally discovered or present with life threatening complications like secondary infection, rupture with haemorrhage and volvulus or intestinal obstruction.⁸⁻¹⁰

Ultrasonography reveals a cystic lesion with multiple thin septa (honeycomb or cobweb pattern) and aspiration of the cystic contents yields a chylous aspirate. On computed tomography (CT), they appear as uni- or multilocular masses with enhancement of the wall and

septum by contrast medium.¹¹ Magnetic resonance imaging (MRI) is the most useful radiological tool. It helps in differentiating from mesenteric cyst by providing good differentiation of cystic and septal structures. Also cystic lymphangiomas lack demonstrable fat content by chemical shift and fat saturation, as clearly seen in the MRI of dermoid cyst.¹²

The definitive diagnosis of lymphangioma is based on histopathology and immunochemistry: the lining mesothelial cells are immunoreactive for cytokeratin and negative for factor VIIIIs. Double staining with Prox1 and CD31 is the most reliable method for characterizing lymphangioma endothelial cells.¹³

Lymphangiomas are classified as simple, cavernous and cystic. The simple type is mostly situated superficially in the skin and composed of small thin walled vessels, while the cavernous variety consists of dilated lymphatic vessels with patent communication with normal adjacent lymphatics. Cystic lymphangiomas is composed of large lymphatic spaces surrounded by collagen and smooth muscles and do not have communication with adjacent normal lymphatics.¹⁴

Primary treatment of mesenteric cystic lymphangiomas is radical surgical excision even when asymptomatic for its potential to grow enormously to invade adjacent structures, develop complications and its risk of sarcoma transformation on irradiation.^{15,16} However, radical excision can sometimes be technically impossible due to its infiltration of vital structures.¹⁷

CONCLUSION

Mesenteric cystic lymphangiomas are rare lesions. It is an uncommon benign tumour of congenital origin. It presents either with chronic abdominal pain or acute life threatening complications. It is usually detected incidentally by radiological studies done for other reasons. Occasionally diagnosis is made during surgery. When detected radical excision should be the practise even when asymptomatic to avoid future presentation with life-threatening conditions.

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