British Journal of Medicine & Medical Research
13(6): 1-4, 2016, Article no.BJMMR.23864
ISSN: 2231-0614, NLM ID: 101570965

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Macrocystic Lymphatic Malformation: A Rare Cause of Acute Abdomen

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Authors' contributions

This work was carried out in collaboration between all authors. Authors KAA and MM were the surgeons who operated on the patient and further managed him. Authors LDK and PAK were the pathologists who reported the case. Authors LDK and ABA did the literature and manuscript. The final work was reviewed by all authors.

Article Information

DOI: 10.9734/BJMMR/2016/23864
Editor(s):
(1) Fuhong Su, ICU Laboratory, Erasme Hospital, Free University Brussels, Brussels, Belgium.
Reviewers:
(1) Tariq Abbas, Hamad General Hospital, Doha, Qatar.
(2) Cosimo Sperti, University of Padua, Italy.
(3) Oguzhan Guven Gumustas, Acıbadem Hospital, Turkey.
Complete Peer review History: http://sciencedomain.org/review-history/13109

Case Study

Received 26th December 2015
Accepted 18th January 2016
Published 28th January 2016

ABSTRACT

Lymphatic malformation (lymphangioma) is an uncommon benign vascular tumour with unknown etiology, which is rarely seen in the abdomen. Clinically it may present with acute abdominal pain requiring surgery and may mimic many other intrabdominal emergencies. A case of a mesenteric macrocystic lymphatic malformation presenting with signs and symptoms of ruptured appendicitis in a 13 year old boy is presented. Post operatively a histopathological diagnosis of macrocystic lymphatic malformation was made.

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Keywords: Mesenteric; macrocystic lymphatic malformation; acute abdomen.

1. INTRODUCTION

Lymphatic malformation (Lymphangioma) is an uncommon benign vascular tumour with unknown etiology. It usually occurs in the head, neck and axillary region of the body [1-3]. Within the abdominal cavity, lymphangiomas are rarely seen. However if they occur, the commonest site of occurrence is the small bowel mesentery, omentum, mesocolon and retroperitoneum [3,4].

Lymphatic malformations are more common in males than females with a ratio of 5:2 and the mean age of presentation is 2 years. Clinically, lymphangioma have numerous presentations including acute abdomen. They may also be asymptomatic [3-6].

2. CASE REPORT

A 13 year old boy was referred from a local hospital to the Cape Coast Teaching Hospital after presenting with a day's duration of colicky abdominal pain that started in the peri-umbilical region. The pain later became constant and localized in the right iliac fossa. He was febrile but did not complain of nausea, vomiting, diarrhea or constipation. Examination showed an adolescent boy with a temperature of 37.6℃, he was neither pale nor icteric and was well hydrated. His abdomen was tender in the right iliac fossa and suprapubic region with rebound tenderness and guarding his bowel sounds were present and normal. There were no other significant systemic findings. A Full blood count showed mild leukocytosis (12.63x10^9), consisting predominantly of neutrophils (9.08x10^9). Abdominal ultrasound scan revealed a hyperechoic mass within the pelvis with lattice-like pattern and a volume of 727.2 cm. A final clinical preoperative diagnosis of ruptured appendicitis with peritonitis was made and patient sent for surgery. At surgery, a mass was found in the mesentery of the jejunum. The mass with a part of the jejunum were resected with end to end anastomosis of the jejunum. Post operatively, patient was on ciprofloxacin and metronidazole and was discharge on post-operative day five.

At gross pathological examination, the mesenteric mass measured 10x9 cm and was lobulated, its surface was covered with fibrino-purulent exudate. Cut surface of the mass showed multiloculated cystic areas containing brown fluid. There was no connection between the mesenteric mass and the lumen of the bowel. The mucosa and serosa of the bowel were grossly normal. Histopathology showed a multiloculated cyst composed of dilated lymphatic channels of varying sizes and shape. The channels contained amorphous material with cholesterol clefts, lymphocytes and hemosiderin laden macrophages (Fig. 1). In some areas of the stroma, there were aggregates of lymphocytes, plasma cells and neutrophils in a myxoid background. There was no malignancy. A histopathological diagnosis of lymphangioma was made.

The patient recovered from surgery and is continuing his education.

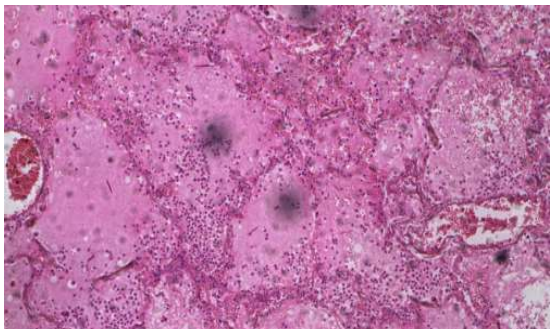

Fig. 1. Micrograph showing the interconnected lymphatic channels with scattered lymphocytes

3. DISCUSSION

Abdominal macrocystic lymphatic malformation is a rare benign congenital malformation of the lymphatic channels forming about 5-6% of all benign tumours in infants and children. The birth incidence of intra-abdominal macrocystic lymphatic malformation ranges from 1/100000 to 1/250000 [7-9]. Forty percent of lymphatic malformation present by age, 1 year and 80% by age of 5 years [3,6,10]. Although most cases of lymphatic malformation present in early childhood our patient was an adolescent with mesenteric lymphatic malformation. Lymphatic malformation usually occurs in the head and neck region although it can occur in any body region. In the abdominal cavity the commonest site is the mesentery. But it can also occur in the omentum, retroperitoneum and mesocolon.

There are a number of theories explaining the occurrence of lymphatic malformation. The main theory explains lymphatic malformation as a proliferation and dilatation of blind ended lymphatic channels during foetal development forming cystic spaces lined by endothelial cells that lack communication with small bowel lymphatics and main systemic lymphatics. This theory may be supported by the fact that most cases presents before age 1 year. Other notable potential causes are abdominal trauma, localized lymphatic degeneration and lymphatic obstruction [3,10,11]. There was no such history suggesting this underlying cause and therefore we attributed it cause to congenital malformation.

Although abdominal cystic lymphatic malformation may be asymptomatic, it may present with symptoms ranging from increased abdominal girth, sensation of abdominal fullness to vague abdominal pain. Acute pain can be due to torsion or haemorrhage into the cyst or intestinal obstruction [7]. In a rare case, Akwei et al. presented a mesenteric lymphatic malformation presenting with signs and symptoms of acute pancreatitis [7]. Our patient presented with signs and symptoms of acute abdomen which preoperatively was mimicking a ruptured appendicitis but intraoperatively the appendix was found to be normal.

The diagnostic procedure of choice in cases of suspected abdominal cystic lymphatic maformation is abdominal ultrasound. Ultrasonographic features of lymphatic malformation are characteristic multiloculated cystic mass with septae of variable thickness [12,13]. In the series done by Kapoor et al. ultrasonographic features described included; cysts with thin or thick septae and varying proportions of solid areas [12]. The ultrasound findings of the lesion in our patient showed cysts with thin septae with a lattice appearance which was confirmed on gross histopathological examination. Although the radiological description was pointing to lymphatic malformation, the conclusion by the radiologist was a ruptured appendicitis due to the clinical presentation mimicking appendicitis. This buttresses the fact that, mesenteric lymphatic malformation may be missed unless there is a high index of suspicion.

Complete surgical excision as was done in our case is the treatment of choice for abdominal cystic lymphangioma associated bowel resection as in the index case is a result of the attachment of the small intestine with the lymphangioma [13].

4. CONCLUSION

Mesenteric lymphatic malformation, a rare benign vascular tumour with unknown aetiology which usually presents in childhood as an asymptomatic mass with increasing abdominal girth may present in adolescence with features mimicking acute appendicitis.

CONSENT

Consent for this publication was obtained from the patient and his guardian.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
http://sciencedomain.org/review-history/13109