

Case report

Benign cystic lymphangioma of sigmoid mesocolon presenting as abdominal catastrophe: an extremely rare entity

S Sah¹, S Nawal¹, R Gupta¹, S Kafle²

¹ Department of Surgery & ²Department of Pathology B.P. Koirala Institute of Health Sciences, Dharan, Nepal

Abstract

Mesenteric cystic lymphangiomas (MCLs) are uncommon benign cystic tumours of unknown aetiology, most often seen in paediatric patients. The clinical presentation ranges from an incidentally discovered abdominal cyst to symptoms of acute abdomen. A 10-year-old male presented with generalized abdominal pain, nausea and vomiting & constipation of several hours duration. Emergency laparotomy revealed multiple cystic masses along both the sides of sigmoid colon and mesocolon, which were inflamed mimicking diverticulosis. Sigmoidectomy with colorectal anastomosis was performed. The postoperative course was uncomplicated. MCLs should be included in the differential diagnosis of cystic intra-abdominal lesions. Even when asymptomatic and discovered incidentally, they must be treated surgically because of the potential to grow, invade vital structures and develop life-threatening complications.

Keywords: cystic lymphangioma, intestinal obstruction, mesenteric,

Introduction

Mesenteric cystic lymphangiomas are cystic lesions of unknown etiology most commonly seen in paediatric patients. Abdominal cystic lymphangiomas are rare congenital benign malformations of the lymphatic system commonly located in the small-bowel mesentery, followed by the greater omentum, mesocolon and retroperitoneum.¹

Lymphangiomas are commonly found in the head and neck region; but intraperitoneal and retroperitoneal locations are very unusual. Abdominal cystic lymphangiomas are more common in boys with mean age of presentation at 2 years.²

The diversity of clinical presentations is seen ranging from incidentally found abdominal cyst

on scans to the symptoms of acute abdomen. We here report an unusual case of 10-year-old boy who presented with mesenteric cystic lymphangioma of sigmoid mesocolon with acute bowel obstruction.

Case report

A 10-year-old boy presented in the emergency department with generalized abdominal pain, diffuse abdominal distension, nausea and vomiting and obstipation of 48 hours duration. He had dull aching abdominal pain and discomfort on and off for previous one month. His vitals were stable. Physical examination revealed diffuse abdominal distension and tenderness with a palpable cystic mass in left lower abdominal region. Bowel sounds were exaggerated.

His blood investigations reports were within normal range. Plain abdominal radiograph showed dilated bowel loops with multiple air fluid levels suggesting bowel obstruction.

Address for correspondence

Dr. Suresh Prasad Sah

Assistant Professor, Department of surgery

B P Koirala Institute of Health Sciences, Dharan, Nepal

Tel: 00977-9842053013

Email: Sureshsah214@gmail.com

Ultrasonography demonstrated dilated bowel loops with hypoechoic cystic lesion of approximately 14x 12x10 cm size in left lower abdominal cavity suggestive of mesenteric cyst.

We suspected mesenteric cyst presenting as acute bowel obstruction and planned for surgical intervention in emergency. Emergency laparotomy was performed which revealed multiple cystic masses along both the sides of sigmoid colon and mesocolon, which were inflamed mimicking diverticulosis which are shown in figure 1 and 2. Sigmoidectomy with colorectal anastomosis was performed and thorough peritoneal lavage was done. Resected sigmoid colon is shown in figure 3. Histopathology showed serosal outpouching with presence of multiple ectatic channels filled with eosinophilic, proteinaceous material along with lymphocytes and the intervening stroma showed dense lymphoid aggregates with few muscle bundles which is shown in figure 4. The overall features were suggestive of mesenteric cystic lymphangioma.

The postoperative course was smooth and uneventful. Patient was discharged on fifth postoperative day. No recurrence of disease was noted in 9 month follow up.

Discussion

Overall lymphangiomas account for about 5-6% of all benign tumors in children.^{1,3} About fifty percent of cases involve head & neck region with only 10% occurring in intra-abdominal region.² Sixty percent of these intra-abdominal cystic lymphangiomas are found at the time of birth. Abdominal cystic lymphangiomas are very uncommon entity. Almost ninety percent are detected by the mean age of two years and most commonly arise from the small bowel mesentery followed by greater omentum.⁴ Large bowel mesocolon is third commonest site of origin in the intra-abdominal region followed by retroperitoneum.

The etiology for abdominal cystic lymphangioma is unknown but the proposed hypothesis for

this rare entity is an embryological failure of the fusion of the lymphatic system: lack of communication between the bowel lymphatic tissue and main lymphatic vessels during fetal development resulting in blind cystic lymphatics lined by endothelial cells.

The diversity of clinical presentations is seen ranging from incidentally found abdominal cyst to the symptoms of acute abdomen. The symptomatology depends on size and location of cyst. Abdominal discomfort is common symptom as seen in this patient but may present with acute peritoneal symptoms due to rupture, volvulus, hemorrhage or infection occurring infrequently.

Abdominal ultrasonography is imaging modality of choice when abdominal cystic lymphangioma is suspected even during antenatal period. On ultrasonography it is seen as a well circumscribed cystic anechoic lesion with septations. Computerized tomography scan may be required in doubtful cases to know the exact location and extension. It appears as smooth unilocular or multilocular cystic masses with homogenous fluid attenuation.

The confirmative diagnosis is always made on the histopathological findings, since this examination shows extensive myofibroblastic involvement and identifies the lymphatic character of it. The differential diagnoses of abdominal cystic lymphangiomas are other fluid filled lesions like pseudocysts, dermoid cysts, enteric duplications and lymphocoele.⁵

The definitive modality of treatment is surgery; it consists of enucleation if feasible.⁶ The segmental resection of bowel is often required because of the close relation between the cyst and the intestinal wall as was the case with our patient. Some authors recommend conservative treatment in asymptomatic patients as there is 10% chance of spontaneous regression. The sclerotherapy technique using doxycycline is alternative but there is high recurrence rate.^{2,5} Laparoscopic resection can also be done, but

the recurrence rate has been reported up to 10% due to incomplete resection.⁷

Conclusion

The abdominal cystic lymphangiomas, rare congenital benign lesions of lymphatic system are frequently seen in paediatric patients and are asymptomatic, usually discovered incidentally on scan, must be treated surgically because of its potential to grow, invade vital structures and develop life-threatening complications. To prevent recurrence complete resection of abdominal cystic lymphangiomas with or without intestinal resection is mandatory.

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