Other full case

Giant cystic lymphangioma in childhood: a rare differential for the acute abdomen

Sabrina Talukdar,¹ Swethan Alagaratnam,¹ Ashish Sinha,² Christopher Charles Thorn,¹ Colin Elton¹¹Department of General Surgery, Barnet and Chase Farm NHS Trust, London, UK;²Polypsis Registry, St. Mark's Hospital, London, UK**Correspondence to** Christopher Charles Thorn, christhorn77@yahoo.com**Summary**

Cystic lymphangiomas are rare benign tumours of childhood resulting from an abnormal development of the lymphatic system, most commonly arising in the head and axillary region. We report a case of haemorrhagic intra-abdominal cystic lymphangioma presenting as an acute abdomen. A 5-year-old girl was admitted with low-grade fever, generalised abdominal pain and elevated inflammatory markers, and a clinical diagnosis of acute appendicitis was made. At operation, two large fluid-filled haemorrhagic cystic lesions were found to occupy most of the abdominal cavity. The lesions were completely excised and histological examination identified them as cystic lymphangiomas. This case report and literature review highlights aspects of the presentation which might have resulted in a preoperative diagnosis, which is seldom achieved.

BACKGROUND

The case describes an unusual pathology and aims to both interest the reader and to re-assert important clinical lessons. The patient's presentation was consistent with acute appendicitis and she was promptly taken to the operating theatre. However, features pointing to the correct diagnosis were available from the history (possible mass), examination (distension relatively early in natural history) and plain x-ray (unusual gas pattern). As well as describing a complication of any intra-abdominal cyst, the report serves to reiterate important clinical principles.

CASE PRESENTATION

A 5-year-old girl was admitted with a 24-h history of worsening central abdominal pain migrating to the right iliac fossa, associated with anorexia and bilious vomiting. A similar but self-limiting episode occurred 3 days prior to admission. She had no significant medical or surgical history and was born at full term by uncomplicated vaginal delivery. She attained normal developmental milestones and had been fully compliant with the standard immunisation schedule.

Initial clinical examination revealed a quiet, dehydrated and unwell child with a regular pulse of 120 bpm, blood pressure 110/62 mm Hg, respiratory rate 18/min and temperature 37.5°C. The ENT and respiratory examinations were normal. The abdomen was distended with localised peritonism in the right iliac fossa, as evidenced by percussion tenderness with interobserver agreement. The consensus clinical impression was that the patient most likely was with acute perforated appendicitis. Following the acquisition of informed consent and within 5 h of presentation, the patient proceeded to the operating theatre.

INVESTIGATIONS

Index blood results revealed neutrophil leucocytosis of $22.3 \times 10^9/l$ and C reactive protein of 237 mg/l to be the only

abnormalities. A plain abdominal radiograph was acquired in view of the abdominal distension. This demonstrated an abnormal bowel gas pattern although its significance was not appreciated preoperatively (figure 1).

TREATMENT

After general anaesthesia and muscle relaxation, the abdomen was again palpated but no discrete mass perceived. Diagnostic laparoscopy was considered but not undertaken due to marked abdominal distension and associated diaphragmatic splinting. A modified Lanz incision and muscle splitting approach was performed. On entering the peritoneal cavity, no appendix was identified and a large darkly coloured mass was encountered. A low, midline laparotomy was required to explore the abdomen sufficiently. The primary pathology comprised two large cysts, arising within the greater omentum. No macroscopic abnormality was identified in the remaining abdominal viscera including the appendix. The smaller cyst, measuring approximately 10×5 cm, was aspirated of 40 ml of blood-stained fluid. Both cysts were excised in their entirety with maximal preservation of the omentum (figure 2). The appendix was also removed. The child was referred to a tertiary paediatric unit for postoperative management where she made an uncomplicated recovery and was discharged on the fourth postoperative day. When further history was taken retrospectively and in the light of the operative findings, the parents mentioned their suspicion of an intra-abdominal mass over several years, although this had never been formally investigated, or highlighted at presentation.

OUTCOME AND FOLLOW-UP

The lesions were measured at 140×95×45 mm (approximately 300 cc) and 95×55×15 mm. Histopathological examination confirmed cystic structures containing blood clot. The cyst wall demonstrated an attenuated endothelial lining with underlying smooth muscle, surrounded by

Figure 1 Abdominal radiograph demonstrates non-distended small bowel loops in the left upper quadrant with a ground-glass appearance in the right lower quadrant.

fibroadipose tissue with moderate active chronic inflammation. Markedly dilated lymphatic channels with adjacent small aggregates of lymphocytes were reported. These appearances were diagnostic of cystic lymphangioma complicated by acute haemorrhage. The appendix was not inflamed and contained *Enterobius vermicularis* within the lumen.

DISCUSSION

Intra-abdominal cystic lymphangiomata are rare congenital benign tumours, where lymphatic channels in the small bowel mesentery or retroperitoneum do not communicate with the main vessels, resulting in sequestration and

formation of cysts. Ninety-five per cent of such lesions occur in the head and axillary region in locations of subcutaneous primitive lymph sacs where they are often referred to as 'cystic hygromas'. They account for between 1 in 27 000 and 1 in 250 000 hospital admissions and occur mainly in children.¹ A recent study series showed that they usually affect boys, with a male-to-female ratio of 5:2 and a mean age of presentation of 2.2 years.² These lesions can be associated with specific genetic abnormalities, most notably Turner syndrome.³ Intra-abdominal cystic lymphangioma is particularly rare, accounting for 2–8% of all cases and the symptoms are variable.⁴ In one series of 22 cases, one-half the cases had an asymptomatic

Figure 2 Cystic lymphangioma at surgery situated within the greater omentum and demonstrating a dark discolouration related to recent intracystic haemorrhage.

abdominal mass while the remainder typically presented with chronic abdominal pain with or without distension.⁵ Less commonly, they can present as an 'acute abdomen', characterised by acute abdominal pain, distension, vomiting, fever and peritonitis.² Ascites may be incorrectly diagnosed when some cysts are large.⁵

Plain abdominal radiographs most commonly reveal a soft-tissue mass with displacement of bowel loops but may identify complications such as intestinal obstruction.² Abdominal ultrasonography is the imaging modality of choice, revealing a sharply defined cystic or multicystic mass often with internal septations. Acute cases with intracystic haemorrhage can be more difficult to diagnose and CT can be more sensitive in providing additional preoperative information including precise anatomical location, adjacent organ involvement, size and complications.⁶

Although an abdominal lymphangioma is considered benign, it may become locally invasive. Complete surgical resection is the treatment of choice, especially if there are signs that the mass is enlarging in size or producing acute symptoms.² Simple aspiration is associated with a high recurrence rate.⁵ If there is bowel involvement, a segmental intestinal resection may be required. In rare cases where the malformation is diffuse, extensive resection may result in short bowel syndrome.⁷

Prognosis following complete resection is excellent. There is a 10% postoperative recurrence rate if resection is incomplete and in such cases, long-term surveillance has been advocated.⁵⁷ Malignant change or spontaneous regression has also been reported.⁸ Presentation with life-threatening haemoperitoneum as a result of spontaneous haemorrhage has also been reported.⁹

In conclusion, intra-abdominal cystic lymphangioma may present with an acute abdomen, due to intra-cystic

haemorrhage, typically in a young child. Children who present with complications of previously asymptomatic congenital problems pose a diagnostic challenge. In this case, the diagnosis may have been elicited from a more extensive history, detailing the parents concerns regarding chronic abdominal distension, unusual examination features and an abnormal bowel gas pattern on abdominal x-ray.

Learning points

- ▶ Congenital abnormalities present outside the neonatal period as surgical disease.
- ▶ Intra-abdominal cystic hygroma is an established differential diagnosis in the paediatric acute abdomen.
- ▶ Confirmation bias affects all doctors and several incongruities were overlooked in this case out of concern for the patient's presumed acute appendicitis.
- ▶ Abdominal distension is a rare presenting feature of acute appendicitis.
- ▶ Abnormal gas pattern may be caused by displacement, suggesting intra-abdominal mass.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Hardin WJ**, Hardy JD. Mesenteric cysts. *Am J Surg* 1970;**119**:640–5.
2. **Konen O**, Rathaus V, Dlugy E, *et al.* Childhood abdominal cystic lymphangioma. *Pediatr Radiol* 2002;**32**:88–94.
3. **Rekhi BM**, Esselstyn CB Jr, Levy I, *et al.* Retroperitoneal cystic lymphangioma. Report of two cases and review of the literature. *Cleve Clin Q* 1972;**39**:125–8.

4. **Fernández Hurtado I**, Bregante J, Mulet Ferragut JF, *et al.* Abdominal cystic lymphangioma. *Cir Pediatr* 1998;**11**:171–3.
5. **Hebra A**, Brown MF, McGeehin KM, *et al.* Mesenteric, omental, and retroperitoneal cysts in children: a clinical study of 22 cases. *South Med J* 1993;**86**:173–6.
6. **Davidson AJ**, Hartman DS. Lymphangioma of the retroperitoneum: CT and sonographic characteristic. *Radiology* 1990;**175**:507–10.
7. **Steyaert H**, Guitard J, Moscovici J, *et al.* Abdominal cystic lymphangioma in children: benign lesions that can have a proliferative course. *J Pediatr Surg* 1996;**31**:677–80.
8. **Henzel JH**, Pories WJ, Burget DE, *et al.* Intra-abdominal lymphangiomata. *Arch Surg* 1966;**93**:304–8.
9. **Safdar A**, Bakhsh M, Ahmed I, *et al.* An unusual cause of haemoperitoneum in a child. *J Pak Med Assoc* 2008;**58**:458–60.

This pdf has been created automatically from the final edited text and images.

Copyright 2011 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Talukdar S, Alagaratnam S, Sinha A, Thorn CC, Elton C. Giant cystic lymphangioma in childhood: a rare differential for the acute abdomen. *BMJ Case Reports* 2011;10.1136/bcr.04.2011.4105, date of publication

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow