

JEJUNAL LYMPHANGIOMA PRESENTING AS AN OVARIAN MASS

JR Norris¹, M Stacey², R S Rampaul³, K L Cheung³

¹16 Close Medical Support Regiment (CMSR), now StR in Surgery. ²16 CSMR now StR in Medicine. ³Department of Surgery, Nottingham City Hospital.

Abstract

Intra-abdominal lymphangiomas are rare benign tumours that usually arise in the mesentery of the small bowel. We present the case of an intra-abdominal lymphangioma that involved both the jejunum and its adjacent mesentery that ultrasound and CT scanning suggested was an ovarian tumour. Laparoscopy confirmed normal ovaries and a jejunal mass which was resected at laparotomy. This circumstance has very rarely been described in the literature and represents a diagnostic dilemma of which clinicians should be aware. Intra-abdominal lymphangioma should be included with other non-gynaecological diseases in the differential diagnosis of a pelvic mass, and requires a multi-specialty approach.

Introduction

Lymphangiomas are uncommon, hamartomatous, congenital malformations of the lymphatic system that involve the skin and subcutaneous tissues, which usually present in childhood. They may occur anywhere in the skin and mucous membranes, with the most common sites being the head and neck, followed by the proximal extremities, the buttocks and trunk. Intra-abdominal lymphangiomas are rare benign lesions that usually arise in the mesentery of the small bowel; they are occasionally found in the intestines themselves or the pancreas. They may present acutely in adult life as a consequence of compressive effects or from their complications and are frequently misdiagnosed preoperatively [1]. We present the case of an intra-abdominal lymphangioma that unusually involved both the jejunum and its mesentery, which was misdiagnosed as an ovarian mass before operation.

Case presentation

A 23 year old woman presented with sudden onset of severe, central abdominal pain and vomiting. There was no history of weight loss, diarrhoea, constipation or urinary symptoms. On examination, she was mildly tender in the epigastrium and lower abdomen, with a palpable, mobile right iliac fossa mass that appeared to arise from the pelvis. She had no history of abdominal surgery. Ultrasonography of the abdomen and pelvis showed a large, complex, solid/cystic mass thought to be ovarian in origin, with free intraperitoneal fluid present. Non-enhanced computed tomographic (CT) scan of the pelvis and abdomen demonstrated a large multi-cystic mass, anterior to the uterus, measuring 9x10.5cm (Figure 1). Her tumour markers were largely unremarkable with a minimally elevated CA125 of 105 with a normal AFP and HCG. A diagnosis of complex right ovarian tumour was made. A further cystic density, adjacent to the left pelvic side wall, was suggestive of either an iliac nodal metastasis or an enlarged left ovary. Multiple loops of dilated small bowel were present. Small soft tissue densities on the surface of several of these loops were suspicious for serosal metastases. During a joint operation

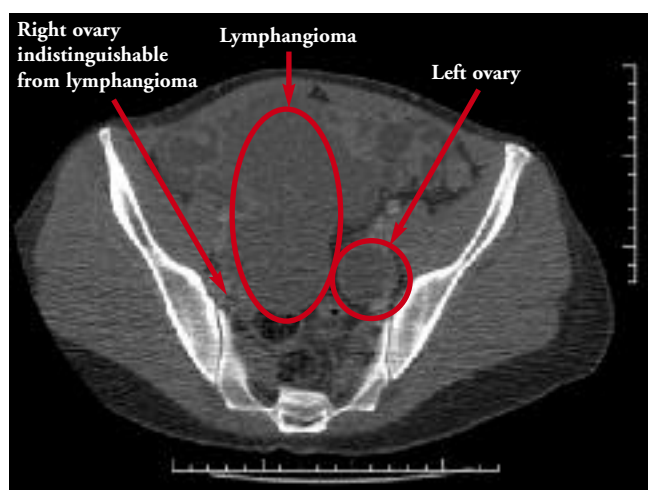


Figure 1: CT scan of a jejuno-mesenteric lymphangioma in the pelvis of a 23yr old woman.

between the general surgeons and the gynaecologists, diagnostic laparoscopy was performed prior to a planned open right salpingo-oophorectomy.

At laparoscopy, a 15x2cm haemorrhagic mass was identified in the pelvis, which appeared to arise from the mesenteric border of a prolapsing loop of jejunum one metre from the duodenal-jejunal flexure. A small amount of intraperitoneal free fluid was present, but there was no evidence of serosal disease and both ovaries appeared normal. After conversion to laparotomy, *en bloc* resection of the jejunal mass and mesentery with end to end anastomosis was performed (Figure 2). The patient made an uneventful recovery and has remained well at surgical follow up.

Cytological examination of the intraperitoneal fluid was non-contributory. The surgical specimen showed a cystic, lobulated, smooth surfaced lesion, up to 10cm in diameter containing numerous blood-filled loculi of up to 2cm diameter. The mass was composed of a proliferation of dilated endothelial lined spaces in a background of fibrous tissue, smooth muscle and adipose tissue. A focal lymphoid infiltrate was present. The mass appeared to arise from the muscularis propria of jejunum and was considered to be an intra-abdominal lymphangioma.

Corresponding Author: Captain Rory Norris, Creag an Roin, Clachan Seil, Oban, Argyll, PA34 4TL

Tel: 07974201428

Email: rory.norris@yahoo.co.uk



Figure 2: Intra-operative image of a jejuno-mesenteric lymphangioma prior to resection and end to end intestinal anastomosis.

Discussion

Intra-abdominal lymphangiomas are rare benign tumours that develop in childhood. It is generally considered that they represent a localized disorder of lymphatic development rather than a truly neoplastic process [2]. Although intra-abdominal lymphangiomas may display infiltrative and aggressive behaviour, there is no evidence of malignant potential. Histologically, they are composed of multiple small or large lymphatic spaces, partitioned by thin walls of fibrous tissue, smooth muscle and lymphoid aggregates. There are three histological categories, two of which, the cavernous and the cystic types, are known to occur within the abdominal cavity. The former is composed of lymphoid stroma and dilated lymph vessels, with connections to adjacent normal lymphatics. The cystic lymphangioma is composed of numerous fluid-filled cysts, which may be serous, chylous, bloody or purulent [3]. This lesion was a cystic lymphangioma.

Abdominal lymphangiomas account for only 5% of all lymphangiomas [4]. They are more commonly found in the mesentery, followed by the omentum, mesocolon and retroperitoneum. They have also been less commonly described in the submucosa of the small and large intestines [3]. The lesion reported here appears to have arisen from the jejunal wall and invaded the mesentery; only 14 previous cases of combined small bowel and mesenteric involvement have been described [3, 5-14].

Intra-abdominal lymphangioma is usually identified as an incidental finding, and for those tumours that manifest in adult life, chronic abdominal pain is the most frequent presentation. Compressive effects and complications account for other modes of presentation, including torsion and rupture of the lesion and intestinal obstruction, volvulus and infarction [3]. The diagnostic problem of differentiating these lesions from other cystic intra-abdominal lesions can be difficult because the imaging features of the lesions overlap [15]. The CT appearance of intra-abdominal lymphangioma is typically that of a cystic, homogenous, low density mass, which may be uni- or multi-locular. Septae of variable thickness may be present. Similar features are demonstrated by ovarian cystadenoma, the most common cause of a pelvic mass in this age group. Only one third of previously described jejuno-mesenteric lymphangiomas were diagnosed pre-operatively, and the treatment of choice is complete surgical resection as it is usually

curative measure with a low incidence of recurrence [16]. Joint management between general surgeons and gynaecologists is essential, both pre- and peri-operatively, whenever there is any significant diagnostic doubt. Most of these masses will turn out to be ovarian in nature and require gynaecological intervention, but diagnostic laparoscopy prior to proceeding to an open operation is recommended to remove any diagnostic doubt and enable the optimal incision to be made.

Conclusion

We present a case of intra-abdominal lymphangioma in a 23 year old woman initially thought to represent ovarian malignancy on cross sectional imaging. Lymphangiomas are frequently misdiagnosed preoperatively despite blood tests, ultrasonography, CT and MRI imaging. These circumstances represent a diagnostic dilemma of which clinicians should be aware. Intra-abdominal lymphangioma should be included with other non-gynaecological diseases in the differential diagnosis of a pelvic mass, or in anyone presenting with acute or chronic abdominal pain due to a mass effect.

Acknowledgements

We would like to thank the following consultant staff of Nottingham City Hospital who contributed to development of the manuscript: Dr T McCulloch (Histopathology), Dr H C Burrell (Radiology), Miss K Williamson (Obstetrics and Gynaecology).

References

1. Takiff H, Calabria R, Yin L and Stabile BE. Mesenteric and intra-abdominal cystic lymphangiomas. *Arch Surg* 1985;**120**:1266-1269.
2. Goldblum JR, Weiss S W. Chapter 26 Tumors of Lymph Vessels. In: Goldblum JR, Weiss S W. *Soft Tissue Tumors*. St Louis, USA: Mosby, 2001:955-83.
3. Seki H, Ueda T et al. Lymphangioma of the jejunum and mesentery presenting with acute abdomen in an adult. *J Gastroenterol* 1998;**33**:107-11.
4. Goh B K P, Tan Y-M, Ong H-S et al. Intra-abdominal and retroperitoneal lymphangiomas in pediatric and adult patients. *World J Surgery* 2005;**29**:837-840.
5. Enomoto H, Yamashita Y, Honda M, et al. Lymphangioma of the ileum and the mesentery of the ileum. Report of a case (in Japanese with English abstract) *Nippon Shoukaki Geka Gakkai Zasshi (Jpn J Gastroenterol Surg)* 1988;**21**:945-948.
6. Campbell W J, Irwin S T, Biggart J D. Benign lymphangioma of the jejunal mesentery: an unusual cause of small bowel obstruction. *Gut* 1991;**32**:1568.
7. Reiker R J, Quentmeier A, Weiss C et al. Cystic lymphangioma of the small-bowel mesentery. *Pathology Oncology Research* 2000;**6**(2):146-148.
8. Mashimo R, Yamazaki T, Hayashi K et al. Lymphangioma of the jejunum: report of a case. *Nippon Geka Hokan - Archiv fur Japanische Chirurgie* 1990;**59**(5):402-407.
9. Candanedo-Gonzalez F, Luna-Perez P. Cystic lymphangioma of the mesentery. Clinical, radiological, and morphological analysis. *Revista de Gastroenterologia de Mexico* 2000;**65**(1):6-10.
10. Nizami S, Mohiuddin K, Daudi I et al. Cavernous transverse mesocolonic lymphangioma in an adult. *Am J Surg* 2007;**193**(6):740-741.
11. Iwabuchi A, Otaka M, Okuyama A et al. Disseminated intra-abdominal cystic lymphangiomatosis with severe intestinal bleeding. A case report. *J Clin Gastroenterology* 1997;**25**(1):383-386.
12. Tsuda K, Murakami T, Nakamura T et al. Lymphangioma of the mesentery and small intestine: a case report showing a solid tumour with a cystic component on US and CT. *Radiat Med* 1994;**12**(5):241-3.
13. Hardin W J, Hardy J D. Mesenteric cysts. *Am J Surg* 1970;**119**:640-645.
14. Chung J H, Suh Y L, Park I A. A pathologic study of Abdominal lymphangiomas. *J Korean Med Sci* 1999;**14**:257-262.
15. Levy A D, Cantisani V, Miettinen M. Abdominal Lymphangiomas: Imaging Features with pathologic correlation. *AJR* 2004;**182**:1487-1491.
16. De Perrot M, Rostan O, Morel P et al. Abdominal lymphangioma in adults and children. *Brit J Surg* 1998;**85**(3):395-397.