

Case Report

Acute presentation of lymphangioma of the retroperitoneum

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CASE REPORT

A 20 year old female student presented to A&E with a one week history of epigastri discomfort radiating to her back with associated nausea and mild constipation. On examination her abdomen was tender but soft. There were no other significant symptoms or signs. She was admitted with a working diagnosis of biliary colic. Routine blood tests were normal. She subsequently underwent abdominal ultrasound and CT imaging.

Abdominal ultrasound demonstrated a large anechoic mass in the upper abdomen postero-inferior and separate from the head of pancreas. However no acoustic 'through enhancement' was seen. There were no gallstones or other significant abnormality. At this stage it was suspected that the lesion was a duplication cyst and CT was performed for further assessment.

CT confirmed the presence of an 8cm by 7cm mass. It was closely related to the duodenum, had

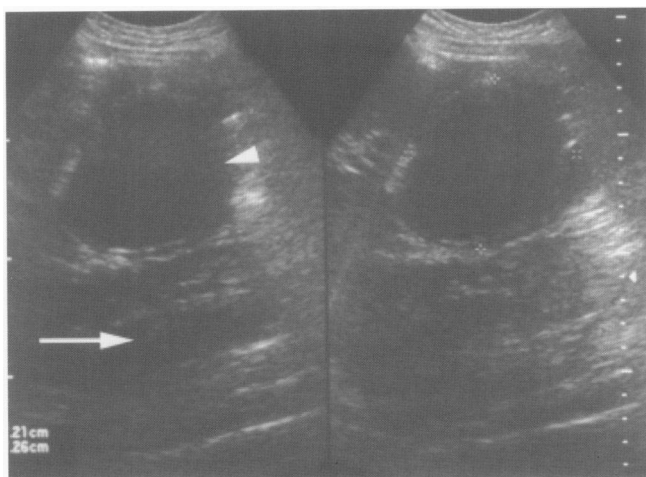


Fig 1. Ultrasound image showing the retroperitoneal lymphangioma (short white arrow). Note the lack of expected 'through transmission' normally expected from a serous cyst (long white arrow).

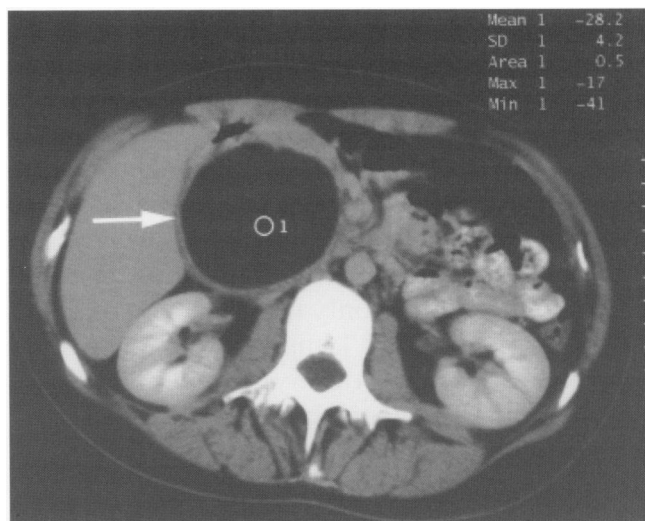


Fig 2. Axial intravenously enhanced CT scan of the upper abdomen with oral radiographic contrast, which shows the large retroperitoneal lymphangioma (short white arrow). An attenuation value has been measured, (less than 0) Hounsfield units (HU), signifying fatty content.

a thick wall and contained homogenous material with an attenuation value of -28 HU i.e. fat density. It did not enhance post administration of intravenous contrast. Differential diagnosis included a lipoma or lymphangioma. An atypical duplication cyst or a large choledochal cyst was also considered. The patient underwent laparotomy and the cyst was resected completely. It contained a 'milky' fluid and a lymphatic vessel was identified entering into it.

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Histopathological examination of the cyst wall demonstrated fibrosis, an inflammatory cell infiltrate, and the presence of endothelial lined vascular channels. These findings confirmed a benign lymphangiomas cyst. The patient remains well.

DISCUSSION

Lymphangiomas are benign lesions of the lymphatic system whose exact aetiology remains uncertain. An acquired abnormality of lymphatic drainage and a congenital lymphatic anomaly are proposed aetiologies, and it is possible both mechanisms are involved. Lymphangiomas are classified by their microstructure into capillary, cavernous and cystic types, although some lesions show a mixture of morphologies, leading to weakness in the classification.¹ The exact morphology of the lesion may depend on the tissues surrounding them. No capillary types have been described in the retroperitoneum. 90% of lymphangiomas occur in patients less than two years old most commonly in the neck or axilla in locations of primitive lymph sacs and are often referred to as 'cystic hygromas', or intra-abdominally as 'omental cysts'. Intra-abdominal and particularly retroperitoneal lymphangiomas are uncommon. Lymphangiomas can occur submucosally and have been described in the stomach and duodenum. In lymphangiomatosis they are numerous and affect many organs including bone. First presentation in adulthood is uncommon, but retroperitoneal cystic lymphangiomas may present incidentally in later life, typically slowly enlarging and remaining asymptomatic for a long period although if large they can present with pressure effects on adjacent organs. If a complication such as infection, haemorrhage, rupture, or torsion occurs presentation can be acute with abdominal pain, and be an unexpected finding at laparotomy. Imaging by USS, CT and MRI is useful in diagnosis and forewarns the surgeon. USS can demonstrate the cystic nature of the lesion which typically appears with sharp margins and scattered internal echoes. Lymphangiomas can be unilocular or multilocular. CT gives a more accurate assessment of relationship to neighbouring organs. Typically at CT, the lesion displaces solid organs, has uniform septa which slightly enhance and has contents of attenuation near that of water. Some groups have found little difference in attenuation readings between serous and chylous-containing lymphangiomas.² In our

case the attenuation of the contents was clearly below that of water. MRI usually demonstrates the typical signal changes of a fluid filled cyst. MRI also can more accurately assess local organ involvement, which can occur with cavernous haemangiomas. Differential diagnoses that should be considered for cystic lesions in the retroperitoneum include duplication cysts, pancreatic pseudocysts, ovarian cysts, retroperitoneal haematomas, retroperitoneal sarcomas, teratomas and abscesses.^{3,4}

Often the diagnosis of lymphangioma can only be confirmed histologically.⁶ Surgical excision is the accepted treatment for symptomatic lymphangiomas and often total excision of the lesion is readily possible with cystic lymphangiomas,¹ however cavernous lymphangiomas are difficult to excise as multiple local adhesions can be present sometimes making excision of a locally involved organ necessary and having high recurrence rates.¹ In some lymphangiomas a stem with a small base can be identified macroscopically. It is important this is excised to prevent local recurrence. If the stem of the cyst and the feeding lymphatics are not ligated chylous ascites will occur.⁵ Recurrence rates for cystic lymphangiomas are low. Rarely do lymphangiomas undergo malignant change or spontaneous regression though both have been documented. Retroperitoneal lymphangioma is an uncommon lesion in adults with the potential to present acutely. Radiological investigation provides important pre-operative diagnostic information essential for effective surgical approach and management.

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