Case Report

Retroperitoneal Lymphangioma

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Lymphangiomas are rare cystic tumours that may present as cystic masses in the retroperitoneum. Retroperitoneal lymphangiomas account for approximately 1% of all lymphangiomas. Confusion with other cystic tumours of the retroperitoneum including those arising from the liver, kidney and pancreas is common. A case of a retroperitoneal cystic lymphangioma occurring in a 41-year-old woman raising interesting diagnostic and management issues is reported. The patient presented with vague abdominal pain and persistent nausea. Radiological imaging demonstrated a large multiloculated thin-walled cyst involving the upper retroperitoneum. Surgical resection was complete, revealing a benign cavernous lymphangioma. The patient made a complete recovery and was disease-free 30 months postoperatively. [*Asian J Surg* 2006;29(1):51–4]

Key Words: cystic tumour, lymphangioma, retroperitoneum

Introduction

An interesting and rare differential diagnosis for a retroperitoneal cystic mass is cystic lymphangioma. A case of a patient presenting with a multicystic mass in the retroperitoneum that was identified as a lymphangioma is reported. These tumours are commonly confused with other cystic masses in the retroperitoneum. The particular diagnostic features and treatment outcomes relating to retroperitoneal lymphangiomas are discussed.

Case report

A 41-year-old woman presented with a 3-month history of vague upper abdominal pain and persistent nausea. She had no previous illness episodes. Colonoscopy and gastroscopy were normal. Full blood count and liver function tests were within normal limits. An ultrasound scan revealed a large cystic mass (8 × 9 × 7 cm) in the region of the porta hepatis (Figure 1A), which was thought to be extrahepatic. Computed tomography (CT) showed a multiloculated cyst possibly arising from outside the liver, compressing the adjacent liver and extending medial to the second part of the duodenum towards the pancreas (Figure 1B). Magnetic resonance imaging (MRI) confirmed a cystic non-enhancing extrahepatic lesion (Figures 1C and D). Carcinoembryonic antigen and CA19-9 were not elevated and hydatid serology was negative.

Preoperative diagnosis based on imaging investigations could not be made with any certainty and a percutaneous biopsy of the lesion was deemed too high-risk due to the location and the possibility of malignant seeding if the lesion was neoplastic.

A laparotomy was ultimately performed. A large retroperitoneal multicystic tumour attached to the uncinate process of the pancreas and coeliac axis and extending to the porta hepatis between the inferior vena cava and portal vein was identified. The cyst was excised intact, with a small portion of attached pancreatic capsule and tissue.

The operation was made technically difficult by the retroperitoneal location of the mass and its intricate relationship with the pancreas and coeliac axis and its tributaries. The thin-walled nature of the cyst rendered it liable to rupture and required handling with extreme care.

Histology

Multiple cystic locules containing thin opaque fluid were

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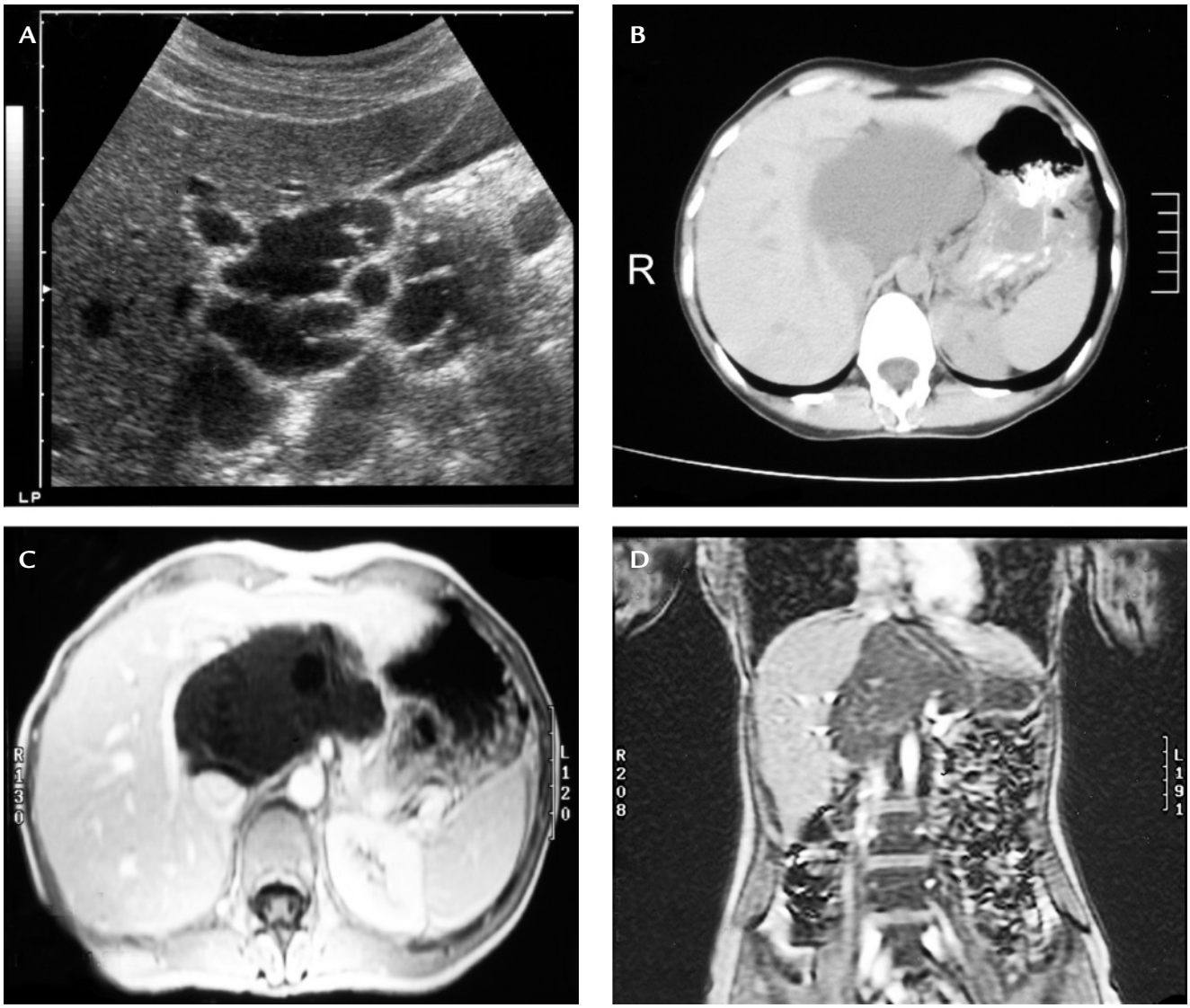

Figure 1. (A) Ultrasound scan showing a complex cystic mass in the region of the porta hepatis. (B) Computed tomography showing a large cystic mass compressing the hilum of the liver. Magnetic resonance imaging with (C) axial and (D) coronal sections showing an extrahepatic cystic mass compressing the porta hepatis, with no gadolinium enhancement and no evidence of a solid component.

noted. The cyst lining comprised trabeculated white tissue with a scattering of firm white and grey nodules within the cyst wall. Histopathology revealed a mass with a central cystic space and a multiloculated appearance, in part lined by a flattened endothelium, with underlying small nodular proliferations of smooth muscle. Large adjoining lymphoid collections were identified (Figure 2). The cyst was adherent in part to a small portion of pancreas tissue. Findings were consistent with a retroperitoneal cavernous lymphangioma.

Follow-up

Recovery was uncomplicated. The patient was symptom-free 30 months postoperatively with no evidence of recurrence on subsequent abdominal imaging.

Discussion

The differential diagnosis of cystic tumour in the retroperitoneum raises several possibilities. These include both malignant and benign tumours. Malignant causes include necrotic neoplasms, germ cell tumours (teratoma), undifferentiated sarcoma, cystic metastases (gastric/ovarian), malignant mesenchymoma, biliary cystadenoma/carcinoma and cystic mesothelioma.[1] Benign cystic lesions of the retroperitoneum include lymphangioma, microcystic pancreatic adenoma and cysts of urothelial and foregut origin. Cysts of foregut origin are subdivided into bronchogenic cysts, which contain cartilage or seromucinous respiratory glands, oesophageal cysts, that are composed of well-developed layers of smooth muscle

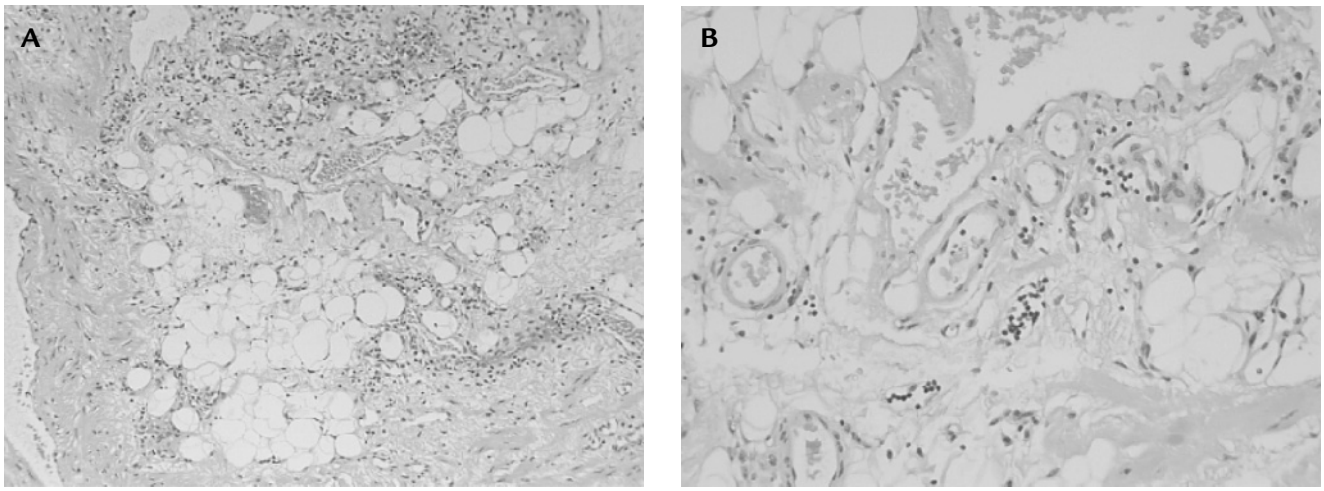

Figure 2. (A) Haematoxylin and eosin section demonstrating the multiloculated appearance of the cyst, with some blood-filled cavities and adjoining lymphoid collections (original magnification × 170). (B) Higher magnification demonstrating a cyst in part lined by flattened epithelium and with underlying small nodular proliferations of smooth muscle (original magnification × 330).

without cartilage, and simple foregut cysts, which have none of these distinguishing features.[2]

In the case presented, the unusual location of the cyst and its intimate relationship with the liver contributes a myriad of other diagnostic possibilities including simple liver cyst, choledochal cyst, biliary cystadenoma/carcinoma, cystic hepatic metastases, hamartomas and parasitic infections such as hydatid disease.[3]

Lymphangiomas are extremely rare benign neoplasms of the lymphatic system with no reports of incidence due to their rarity. They can occur in any location in which lymphatics are normally found. Head and neck lymphangiomas (cystic hygromas) account for 75% of reported cases.[4] The remainder occur in the axillary region (20%) or in other less common locations (5%). Retroperitoneal lymphangiomas account for approximately 1% of all lymphangiomas, with 185 cases identified in a review of international literature.[5]

The aetiology of these benign neoplasms is uncertain, with most authors postulating a combination of inflammatory and fibrotic processes or genetic predisposition. Other factors implicated include mechanical pressure and retention, traumatic factors, degeneration of lymph nodes and disorders of endothelial lymphatic vascular secretion or permeability. The frequent development within areas where primitive lymph sacs occur suggests that lymphangiomas are malformations arising from sequestrations of lymphatic tissue that fail to communicate normally with the lymphatic system.[6] This results in the formation of unicystic or multicystic tumours whose cavity is covered with a layer of endothelium and filled with chylous or serous material.

Intra-abdominal lymphangiomas may arise from the mesentery, greater omentum or retroperitoneum and tend to grow slowly within the abdomen. The majority are asymptomatic and are discovered incidentally on abdominal imaging for other conditions. Symptoms are usually related to tumour size. Approximately 40% of cases present with symptoms of obstruction.[7] Other symptoms include ascites and intermittent fever. Rarely, bleeding or rupture of the tumour will cause an acute abdomen.[5] Clotting disorders have also been described in patients with lymphangiomas.[8] Acute symptoms are more common in children than adults. The average time from occurrence of symptoms to operation is more than 7 months.[9]

In contrast to mesenteric cysts, lymphangiomas not only have the typical characteristics of the lymphatic system, but also have smooth-muscle cells in their walls. Histologically, lymphangiomas are classified as capillary, cavernous or cystic, with only the latter two reported to occur in the retroperitoneal region.

Preoperative diagnosis of lymphangiomas is rare prior to laparotomy or laparoscopy.[5] Abdominal X-rays generally show nonspecific expansion and, in some cases, foci of calcification may be seen. Lymphangiography is seldom used but is sometimes useful in preoperative diagnosis. CT and MRI features of lymphangiomas have been described. The diagnosis of lymphangioma based on these modalities is generally one of many potential differentials for a multiloculated cystic mass arising retroperitoneally. One of the main features of retroperitoneal lymphangioma is that the mass is generally of water density on CT or MRI.[10] Guided biopsy of the lesion is

often difficult and rarely attempted due to the location of tumours and concerns of potential dissemination of malignancy. When fine-needle biopsy is performed, characteristic abundant lymphocytes may be apparent.[10] The likelihood of preoperative diagnosis is greatest when imaging is combined with biopsy.[11]

Outcomes following complete resection of retroperitoneal lymphangiomas are generally good. Surgery is often required for symptom control or diagnosis.[12] Recurrence of symptoms with incomplete excision is possible. Dissemination in the retroperitoneum is very rare but potentially a fatal complication.[13] Injection of sclerosants such as alcohol and bleomycin into lymphangiomas has been described in the literature in nonsurgical candidates. However, induration of the cyst and infection often complicate these procedures.[14,15]

In conclusion, the differential diagnosis of a retroperitoneal cystic lesion raises several possibilities. Cystic lymphangioma should be considered. Despite being rare, these tumours have an excellent prognosis, with symptom relief and cure achieved with complete surgical excision.

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