Case Report

Total Laparoscopic Excision of Unusually Large Giant Retroperitoneal Cystic Lymphangioma

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Abstract

Retroperitoneal cystic lymphangioma is a rare benign congenital malformation of the retroperitoneal lymphatic which usually presents in infancy, but rarely in adult. Due to this rarity preoperative diagnosis is often difficult. This rare neoplasm must be differentiated from other retroperitoneal masses. Laparoscopic excision of such lesions is associated with less morbidity and faster recovery. We present a case of a 41-year-old male with a retroperitoneal lymphangioma, who was managed by laparoscopic excision.

Key Words: Retroperitoneal, lymphangioma, laparoscopy.

Introduction

Retroperitoneal lymphangiomas are rare, benign tumors which have a variable presentation. They need to be differentiated from other retroperitoneal tumors and imaging plays an important role in the diagnosis and management. Mostly, these tumors are removed by open surgery and there is limited literature on laparoscopic removal of such lesions. We report a case of retroperitoneal lymphangioma, which was laparoscopically excised.

Report of Case

A 41-year-old male presents with complaints of pain in right-lower abdomen and nausea for past 1 month.

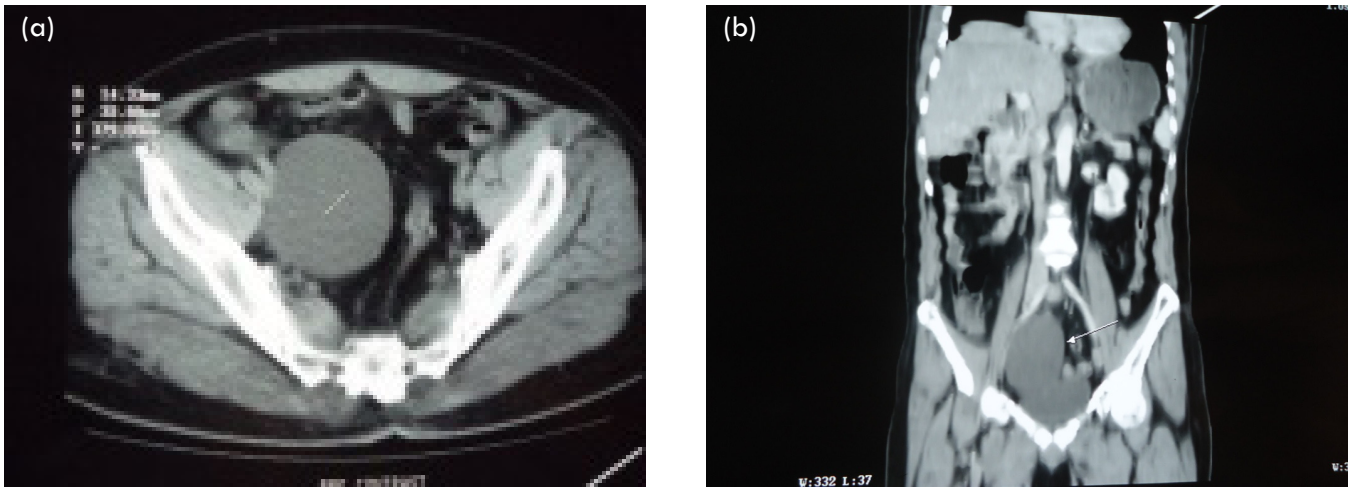

Figure 1: Abdominal computed tomography scan showing large unilocular, hypodense mass seen retroperitoneum: (a) coronal section, (b) sagittal section.

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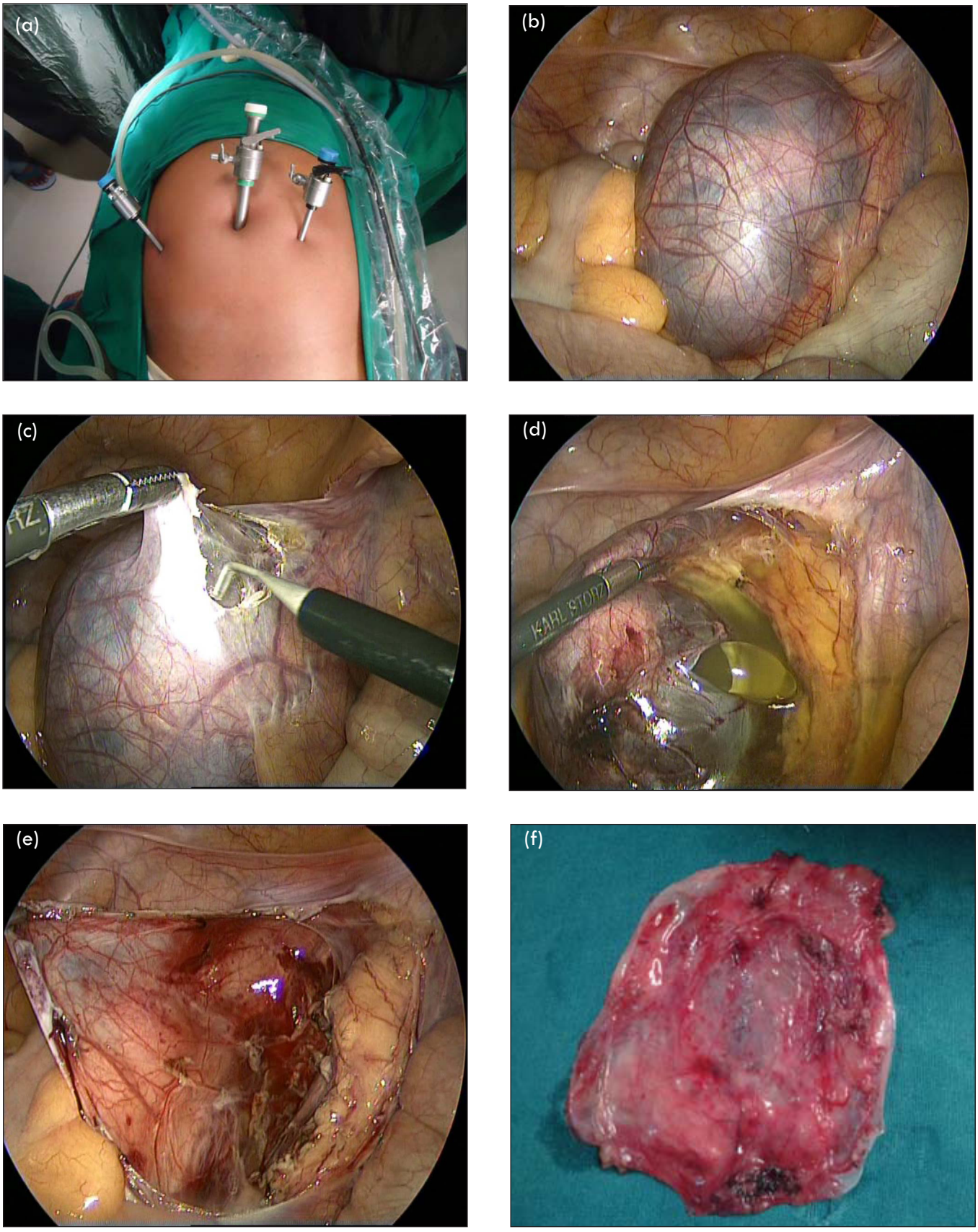


Figure 2: (a) Trocar position, 10 mm umbilical optical trocar, two 5 mm port right and left at level of umbilicus in mid clavicular line. (b) Intraabdominal laproscopic view of cyst. (c-e) Laproscopic excision of cyst. (f) excised cyst.

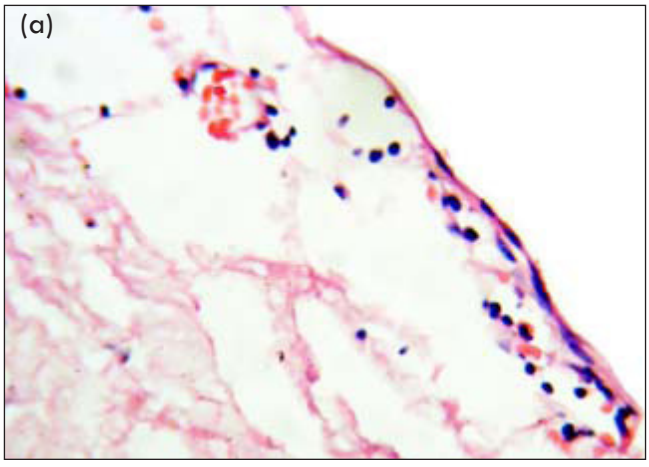
He had associated anorexia, but no loss of weight. He had no urinary or gastro-intestinal symptoms. His physical examination and laboratory studies were normal.

Ultrasound abdomen and pelvis showed single large uniloculated cystic lesion about 6.6 cm x 7.3 cm x 6.7 cm in size, arising from right-lower abdomen and pelvis, anterior-superior to the bladder. The organ of origin of the lesion was not clear on ultrasound. Considering the USG findings, patient was subjected to a CECT abdomen. On computed tomography (CT) scan of abdomen and pelvis (Figure 1 a-b), a well-defined, hypodense lesion with smooth margins was seen in pelvis, in the midline and to right side of abdomen. It measured 8 x 8 cm, and there was no evidence of any enhancement of solid component or septation. The lesion was abutting the urinary bladder inferiorly and vertebral body postero-superiorly. Based on these findings, a differential diagnosis of a lymphangioma, mesenteric cyst or neuoro-enteric cyst was made.

With the preoperative diagnosis of retroperitoneal lymphangioma/mesenteric cyst, the patient was taken for a diagnostic laparoscopy.

Operative Technique

Diagnostic laparoscopy revealed a large, single retroperitoneal, cystic lesion which was separate from the mesentery and had a clear cleavage from all the retroperitoneal organs (Figure 2b). Laparoscopic excision of the mass was carried out via the trans-peritoneal approach by placing 10 mm trocar in umbilicus and 5 mm trocar in right and left mid-clavicular line at the level of umbilicus (Figure 2a).Whole cyst was dissected and retrieved after



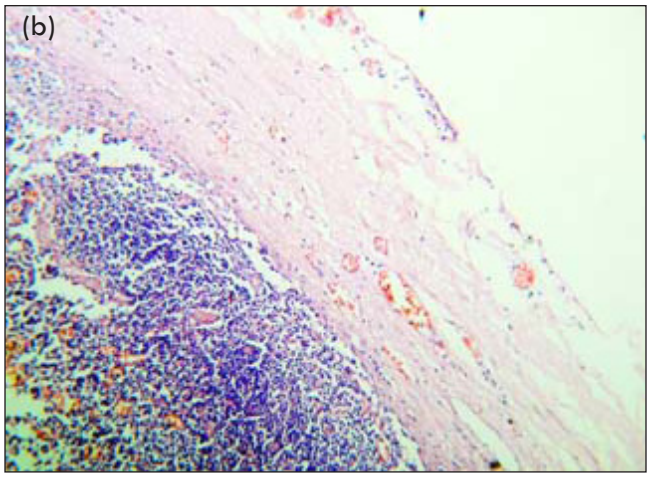


Figure 3: (a-b) Photomicrograph of cystic lymphangioma.

decompression of cystic fluid which was chylous in nature (Figure 2 c-f) through the umbilical trocar. Postoperative period was uneventful and recovery was good. Patient was discharged on postoperative day 2. Histopathological examination confirmed it to be an uncomplicated cystic lymphangioma (Figure 3 a-b). Patient was under regular in follow-ups and has remained asymptomatic till 12 months after surgery.

Discussion

Lymphangioma is a benign congenital malformation of lymphatic channels, which has been classified into two groups: capillary, cavernous, and cystic depending on the size of lymphatic space.[1] Lymphangioma is most commonly found in pediatric patients.[2] The most frequently involved regions are the neck (75%) and axilla (20%).[3,4] Less than 5% of lymphangiomas are diagnosed intraabdominally,[2] and they have been reported in the mesentery, gastrointestinal tract, spleen, liver and pancreas. They are infrequently encountered in the retroperitoneum.

Retroperitoneal lymphangiomas are best described as developmental abnormalities of the lymphatics, and are almost always benign.[5] They are thought to appear due to sequestration of retroperitoneal lymphatic tissue, and lack of its communication with main lymphatic vessels, resulting in formation of cystic masses.[5] Commonly observed retroperitoneal lymphangiomas are usually of cavernous or cystic types of which most reported cases have been of a cystic nature, as was this case.[3] Patients with retroperitoneal lymphangioma are usually asymptomatic and the masses are often found

incidentally by imaging technique or surgery for other purposes.[1] Occasionally, the patient presents with symptoms when the tumor is large, or when infection, hemorrhage, torsion, or rupture occurs.[1] The most common clinical manifestation is that of a slowly enlarging abdominal mass vague abdominal pain, loss of appetite and nausea, as seen in this case. The mass effect is the potential culprit in causing significant morbidity by obstruction (40%) or dislocation of the adjacent organs.

Generally, diagnosis is made by ultrasound and/or CT scan, which are both very sensitive and relatively specific.[6] The final diagnosis is achieved by pathological examination of the specimen after surgical or laparoscopic excision. Various treatment modalities have been described for lymphangiomas. Aspiration followed by sclerotherapy with either OK-432(picibanil) or tetracycline has been illustrated in case reports.[7,8] Sclerotherapy may show excellent results in patients with a single lobe cysts, but repeated and frequent sclerotherapy may be required for patients with multilocular cystic lymphangioma. The use of local argon beam coagulation and sclerotherapy in addition to surgical resection has been described in a case of a life threatening total abdominal lymphangiomatosis.[9]

However, complete surgical excision of cystic lymphangioma still remains the primary treatment of choice. The sacrifice of adjacent structures must be avoided, and in difficult cases surgical and nonsurgical methods may be combined, generally sequentially. Laparoscopic complete excision of retroperitoneal cystic lymphangioma, as in this 41-year-old male patient, has rarely been described in the literature. Tsukamoto and others report a successful case of a complete resection of a retroperitoneal cystic lymphangioma in a 36-year-old woman.[10]

The laparoscopic approach in our patient was technically smooth and feasible. To facilitate precise dissection, we used a monopolar hook. This method was associated with no complication. No recurrence was observed in this patient. The comfort of the patient, with less postoperative pain, rapid recovery after surgery, and little scar formation, is also a considerable advantage associated with this minimally invasive technique. Laparoscopic excision should therefore be considered as a therapeutic option to treat such rare tumor.

Key Points

- Retroperitoneal lymphangiomas are a rare entity.
- These are mainly a diagnosis of exclusion.
- Imaging plays an important role in the diagnosis of these lesions and laparoscopic excision is feasible in these patients.

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