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Laparoscopic treatment of retroperitoneal lymphangioma in an adult: A rare case

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Abstract: Retroperitoneal lymphangiomas are rare lesions and usually asymptomatic and the masses are often found incidentally by imaging technique or surgery for other purposes. Sypmthomatic abdominal lymphangiomas most often present as acute abdomen mimicking intestinal obstruction or appendicitis. Pre-operative diagnosis of retroperitoneal lymphangioma, in general, is challenging and rare, prior to laparatomy or laparoscopy. The diagnosis of lymphangiomas is based on histopathological examination of the mass. Surgery is often required for symptom control or diagnosis and the outcomes of surgery are excellent. Diagnostic tools can not always able to support differential diagnosis in such cases and laparoscopic surgical excision could be safely performed. Herein, we report a 66 year old female patient presented with right upper quadrent pain and underwent surgery to carry out for definitive diagnosis and relief of symptoms.

Keywords: Lymphangioma, Retroperitoneal, Cystic, Laparoscopy

1. Introduction

Lymphangiomas are rare benign vascular lesions and are thought to arise from the abnormal development and/or ectasia of lymphatic vessels or from blockage of proximal lymphatics or trauma(1,2). lymphangiomas mostly (95%) occur in the neck and axillary regions, rarely in the chest and abdominal cavity(3,4). Less than 5% of lymphangiomas are diagnosed intra-abdominally(5). They are infrequently encountered in the retroperitoneum. Retroperitoneal lymphangiomas are rare lesions with incidence of approximately 1%(6). Patients with retroperitoneal lymphangioma are usually asymptomatic and the masses are often found incidentally by imaging technique or surgery for other purposes(7,8). With the advancement of radiographic techniques, the ability to clinically characterize retroperitoneal cystic lesions is improved. But still, the origin of the cyst could still not be distinguished in some cases.

Herein, we report a case of a retroperitoneal lypmhangioma with an emphasis on the symptoms, differential diagnosis and treatment.

2. Case Report

A 66 years old female patient presented with recurrent right flank and right upper quadrent abdominal pain during the last 3 months. On admission, vital signs (blood pressure, heart rate, respiration rate, and body temperature) were within normal limits. The patient was in good general health and had no significant weight loss. On physical examination, the abdomen was soft. The routine blood tests were normal. There was no remarkable family history. Tumor markers were in normal limits.

For evaluation of the right flank pain, abdominal ultrasound was performed revealing a mass measuring around 9.0 × 9.5 cm in size in right upper abdomen. The mass are predominantly anechoic with fine septations. Magnetic resonance imaging(MRI) showed a large homogenous cystic mass measuring 8.5×9.5 cm with well-defined contours, being hypointense on T1-weighted images and hyperintenseon T2-weighted images between the upper pole of the right kidney and liver (Figure 1). The imaging findings favored an unknown originated mass between right kidney and liver. With clinical and radiological findings, we

decided to perform laparoscopic excision of the mass to carry out for definitive diagnosis and relief of symptoms. Laparascopic excision of the mass was performed. The patient recovered from the surgery with no complications. The patient was discharged on postoperative day 3. Surprisingly, the histopathologic examination after the operation revealed grossly the tumoral mass was 10 cm solid, partly hemorragic lesion neighbouring renal tissue with well defined borders. Histologically it was consisted of partly trombosed cystic, thick walled vascular hamartomatous lesions lined by flattened benign cells and lymphoid aggregates were noted on the walls (Fig. 2). Final diagnosis was a pseudotumor composed of trombosed, hemorragic cystic lymhangioma (Fig. 3). After a 6 months follow-up, she remains in good health.

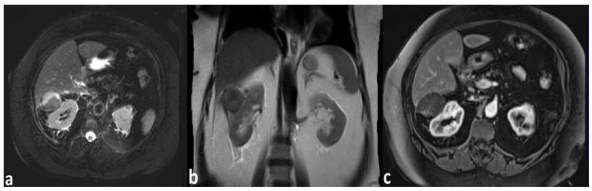

Figure 1. *MRG images. (1a: T2 fat satured transverse MRG image. 1b: T2 coronal MRG image. 1c: T1 fat satured transverse MRG image.)*

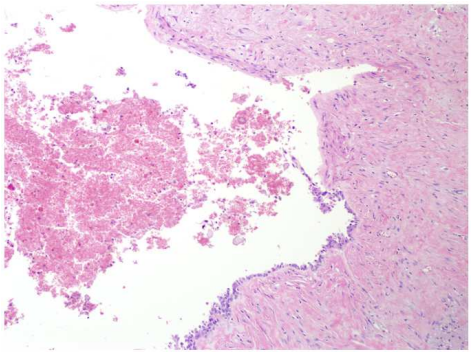

Figure 2. *H&E Cystic, thick walled vascular hamartoma lined by flattened benign cells with some eritrocytes in the lumen.*

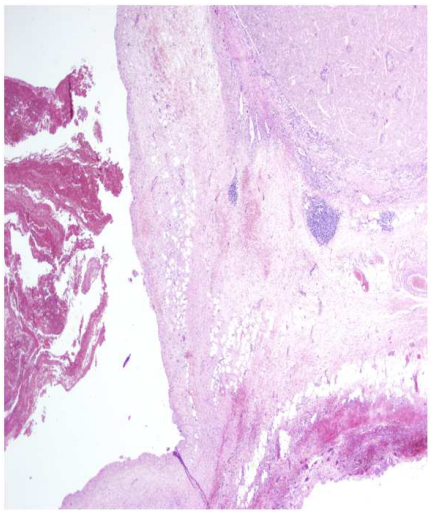

Figure 3. *H&E Thick walled partly trombosed lymphangioma with lymphoid aggregates on the wall.*

3. Discussion

Lymphangiomas are benign vascular lesions that show lymphatic differentiation and mostly (95%) occur in the neck and axillary regions; the remaining 5% are located in the lung, mediastinum, abdominal viscera mesentery and retroperitoneum(3,4). Retroperitoneal lymphangiomas are rare lesions with incidence of approximately 1%(6). Lymphangiomas usually manifest within two years of age, however, retroperitoneal and intra-abdominal lymphangiomas become symptomatic in adulthood due to its slow progressing nature(9,10). The clinical presentation of lymphangioma depends on the anatomic location and tumor size(10,11). Large tumors may present as asysmptomatic masses, most often the masses are often found incidentally by imaging technique or surgery for other purposes(7,8). These tumors may cause symptoms due to compression of adjacent structures leading to a diagnostic consideration of sarcomas(10). Sypmthomatic abdominal lymphangiomas most often present as acute abdomen mimicking intestinal obstruction or appendicitis(8,12).

Cysts of urothelial and foregut origin, microcystic pancreatic adenoma, retroperitoneal hematoma, abscesses, duplication cysts, ovarian cysts and pancreatic pseudocysts, malignant tumors such as cystic mesothelioma, teratoma, undifferentiated sarcoma, cystic metastases and malignant mesenchymoma are differential diagnosis of cystic retroperitoneal lymphangiomas(10). Ultrasonography and CT reveal multiseptate or multilocular cystic masses and have limited roles in the preoperative diagnosis(10). On MR imaging, lymphangioma displays homogenous hyperintense signals in T2-weighted images and low intensity signals in T1-weighted images(8). Pre-operative diagnosis of retroperitoneal lymphangioma, in general, is challenging and rare, prior to laparatomy or laparoscopy(9). The diagnosis of lymphangiomas is based on histopathological examination of the mass. It comprises dilated and interconnected vascular channels lined by flattened endothelial cells with intervening connective tissue septae containing smooth muscle fascicles, lymphocytes and adipose tissue(8,10).

Surgery is often required for symptom control or diagnosis(6). Outcomes following complete resection of retroperitoneal lymphangiomas are generally good(13,14). Dissemination in the retroperitoneum is very rare but potentially a fatal complication(6). Some studies suggest internal peritoneal cavity marsupialisation and also aspiration, drainage, and irradiation of the lymphangioma give poor results(15). Due to its potential to grow, invade vital structures and develop life-threatening complications, complete laparoscopic excision should be considered as a therapeutic option to treat retroperitoneal cystic lymphangioma(13,14).

In conclusion, diagnostic tools can not always able to support differential diagnosis in such cases and cystic lymphangiomas should be considered among possible

diagnoses. Laparoscopic complete surgical excision could be safely performed in retroperitoneal lymphangiomas. With complete excision they have very good prognosis, with symptomatic relief and cure achieved with.

4. Consent

All the authors should confirm that the patient has given their informed consent for the case report to be published.

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