Abstract: Lymphangioma in retroperitoneum is a rare mass-like lesion which presented histopathologically with single or multi-cystic type and/or cavernous type; and clinically is often asymptomatic. Imaging studies was not enough to conclude exact diagnosis due to wide distribution of differential diagnosis resulted from different organ origination of tumor including of pancreas, liver, gastrointestinal and urologic system. This report described a 36 years old women complaining of 8 months vague abdominal pain underwent surgery in order to trans-abdominal total resection of a cystic mass-like tumor just located between abdominal aorta and inferior vena cava. Pathologic studies revealed benign multi-cystic lymphangioma with calcified foci. Symptoms completely removed after surgery and in follow-up period.

Keywords: Lymphangioma, Retroperitoneum, Cystic Tumor

1. Introduction

Retroperitoneal tumor lesions have wide spectrum of differential diagnosis whether malignant or benign. There are reasons that making exact diagnose invalid if no major surgical procedure executes [1] including of difficult access to anatomical position, adjacent vital elements and concerns of seeding malignant cells during guided biopsy approach. [2] Computed tomography (CT) and magnetic resonance imaging (MRI) studies were unsatisfactory diagnostic tools because there are many pathologic changes making retroperitoneal multi-cystic mass lesions. [2] Lymphangioma is one of rare benign mass like lesions developing due to lymphatic vessels malformations or blockage [3] even everywhere of the body that lymphatic exist [4] with the most incidence in head and neck (75%) and the least in retroperitoneal space(1%). [1] This report describes a case with retroperitoneal cystic lymphangioma placed between abdominal aorta and inferior vena cava underwent laparotomy and complete excision of the mass.

2. Case Report

A 36 years old woman complaint of 8 months generalized vague abdominal pain with superiority of preumbilical area with no nausea or vomiting. There was no history of previous similar complaints. Patient had controlled hypothyroidism and was under regular daily consumption of 100 microgram tablet of levothyroxin. Complete blood count, liver function tests and related gastrointestinal enzymes were in normal limits. An Ultrasound study showed a 22x24x38 mm cystic mass which placed 25mm superior to inferior mesenteric artery (IMA) between abdominal aorta (AA) and inferior vena cava (IVC) with neither blood flow support nor originating from large vessels of the zone. An abdominopelvic computed tomography (CT) scan revealed a 50x32x30 mm cystic lesion in aortocaval area just superior to umbilical level with calcified foci in tumor wall. CT-angiography findings also demonstrated a well-defined cystic lesion between AA and IVC with no further encroachement to large vessels (Figure 1). Evaluating of tumor markers including of Carcinoembryonic antigen (CEA) and CA-125 were negative. Preoperative imaging studies failed to imply on a net diagnosis. While
percutaneous sampling from the mass was accompanied with both probable seeding of malignant tumor cells and injury to AA or IVC during procedure, eventually, laparotomy approach was considered.

A retroperitoneal 50x50 mm cystic tumor mass placed directly between AA and IVC and attached to AA, IVC, vertebra and other adjacent soft tissue fully excised intact after releasing adhesions to adjacent elements including AA, IVC, anterior vertebral body and soft tissues. Operation was technically with difficulties as retroperitoneal position of the mass and concurrent attachments to vital vascular elements from different sides of the mass.

2.1. Histopathology

Microscopic studies revealed a cystic tissue containing of eosinophilic acellular materials and fluid. Lymphocytic infiltrations with zones of calcification and cholesterol clefts were noted. Scattered fibrotic changes and peripheral adipose tissue were also regarded. Cytologic studies were negative for malignancy and histopathologic findings were compatible with diagnosis of retroperitoneal multi-cystic lymphangioma (Figure 2). Lymphangioma consists of mingled lymph vessels and smooth muscle elements. Tumor cells are plumper and paler than those of usual leiomyoma.

2.2. Follow-up

Postoperative 6 months recovery period was uncomplicated. Wound healing developed successfully. Abdominal pain recurrence was not noted. Postoperative imaging studies revealed no new pathologic changes except for operation-related changes.

3. Discussion

Retroperitoneum is a potential space for demonstrating of benign and malignant cystic mass like tumors. The latter includes of necrotic neoplasm, germ cell tumors, sarcoma, mesothelioma, biliary carcinoma and gastrointestinal or ovarian metastases. [5] Benign tumors include of lymphangioma, pancreatic adenoma, urothelial and foregut cysts whether contains cartilage and smooth muscles or not. [5] Although etiology of development of lymphangiomatic lesions is even unknown but inflammatory and fibrotic changes, genetic predispositions, traumatic events, mechanical pressures and retentions of lymphatics, lymph node degenerations, secretory and permeability disorders of
lymphtheliom, sequestration and blockage of lymphatic tissues which induce to lymphatic flow discontinuation and impairment of lymphatic system communication would result in lymphangioma. [6] Lesions often found superficially in head and neck (75%), axilla (20%) and rarely in other sides (5%) or retroperitoneum (1%). [1, 4] Intra-abdominal lymphangioma could originate from mesenteric tissue, greater omentum and retroperitoneum. It grows slowly. However the lesion is often asymptomatic and discovery of lymphangioma is incidental but depending on the size of tumor, it could manifest by intermittent fever, gastrointestinal or urologic obstruction, coagulation disorders, anemia, infection, volvulus, torsion, bleeding and cyst rupture which induced to acute abdomen. [7] Symptomatic lymphangioma presented symptoms in 90% of cases under 2 years of age with most involvement of head and neck. [2] While retroperitoneal lesions developed in embryonic period and slowly grow, they would almost find in older children or adults. [8] Lymphangioma also could be a manifestation of Klippel-Trenaunay syndrome in rare cases. [3] Histopathologically, lymphangioma defined as simple or capillary type with small thin-walled lymphatic channels, cavernous type with dilated lymphatic channels and cystic type with characteristics of single or multiple cystic mass with septa [2, 7] The two latter types founded in retroperitoneal space. [5] Previous studies showed no calcified retroperitoneal lymphangioma except of two reports. [9] This study also presented a case with retroperitoneal calcified cystic lymphangioma. Applying CT or MRI studies was not enough to distinguish exact diagnosis for retroperitoneal lymphangioma. [2] Therefore, surgery whether with laparotomy or laparoscopic approach is often needed to make true diagnosis and also to control symptoms by total cyst resection without extended excision. [3, 10] However, recurrence is possible if tumor does not completely remove. [8] Furthermore, cellular dissemination rarely occurs and is a fatal complication. [11] Non-surgical treatment including of cyst drainage and sclerosant agents (such as bleomycin and alcohol) injection have been tried previously but was accompanied by remaining induration, cyst infection and recurrence, therefore, was not preferred in comparison with surgical approaches. [7]

4. Conclusion

Retroperitoneal lymphangioma presents as vague abdominal pain, with radiologic finding of mass lesions along with intra-abdominal lymphatic vessels drainage system. It should be considered as a differential diagnosis of retroperitoneal solid space occupying tumors. Surgical resection could be both curative and histopathologically diagnostic. Although recurrence is possible and surgical re-excision of tumor should be execute.

Conflicts of Interest

Authors did not have any conflicts of interest in writing this article.

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References