CASE REPORT

Multiple retroperitoneal cystic lymphangiomas in a pregnant woman: A case report

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ABSTRACT

Cystic lymphangiomas are extremely rare benign vascular neoplasms generally found in childhood. Thought to arise from obstructed lymphatic channels during development, but may also arise secondary to trauma, inflammation, malignancy, and surgery or radiation exposure. They are usually seen in the head and neck or axillary region, and rarely in the abdominal cavity with only 1% occurring in the retroperitoneum. These are typically asymptomatic lesions incidentally identified by computer tomography, magnetic resonance imaging or ultrasound. Complete surgical resection with histologic confirmation of the diagnosis is the treatment of choice.

We present a case of a 27-year-old woman with multiple cystic lymphangiomas in retroperitoneum, discovered incidentally by ultrasound during pregnancy.

Key Words: Cystic lymphangioma, Retroperitoneal, Pregnancy, Rare neoplasm

1. INTRODUCTION

Cystic lymphangiomas are extremely rare benign vascular tumors originating from the lymphatic channels, and arising in different organs.[1] Typically, childhood neoplasms, 90% of them are diagnosed before 2 years of age.[2] They are usually seen in the head and neck or axillary region (95%); however, they may rarely be seen in the abdominal cavity of adults, with only 1% occurring in the retroperitoneum.[1,2,4] These lymphangiomas are thought to occur secondary to congenital lymphatic obstruction during embryological development that results in lymphangiectasis, they may also arise in adults due to inflammation, abdominal trauma, surgery, radiation exposure or malignancy.[3,4] During pregnancy, they may grow in size due to excessive production of cytokines and vascular endothelial growth factor.[4] When symptomatic, the clinical presentation includes abdominal pain and distension. Some complications include intracystic bleeding, infection, and rupture or compression of adjacent organs. The useful diagnostic techniques are magnetic resonance imaging (MRI), computed tomography (CT) and endoscopic ultrasound with fine needle aspiration;[3] however, a definitive diagnosis requires histologic confirmation after surgery.

We present a case of a young pregnant woman with multiple retroperitoneal cystic lymphangiomas.

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2. Case presentation

We reported an interesting case of a 32.2 weeks young pregnant, who presented with unexplained intense abdominal pain in the left hypochondrium for 24 hours.

An abdominal ultrasound was performed where a complex, large-volume cystic mass occupying the entire left hypochondrium was identified. The mass showed regular edges, peripheral vascularization, and an approximate volume of 1000 mL which appeared to be adjacent to the left ovary.

The patient presented with spiking fever and complete blood cell count demonstrated leukocytosis with a left shift. Microbiology studies of amniotic fluid was negative for microorganisms. The electronic fetal monitoring demonstrated fetal bradycardia, therefore the decision to terminate the gestation was made.

Exploratory laparotomy identified three retroperitoneal cystic lesions, the largest being 14.0 cm in greatest dimension and showed attachment to the splenic vein, and close proximity to the lesser curvature of the stomach, the spleen and pancreas. The smaller lesions each measured 8.0 and 6.0 cm in greatest dimension and were attached to the inferior aspect of the liver and the head of the pancreas, respectively.

These lesions were submitted to pathology for histologic examination. Grossly, three large cystic masses with tan-white smooth translucent surfaces were identified (see Figure 1). Sectioning revealed multiloculated cystic structures filled with serous fluid and areas of fibrosis and calcifications.

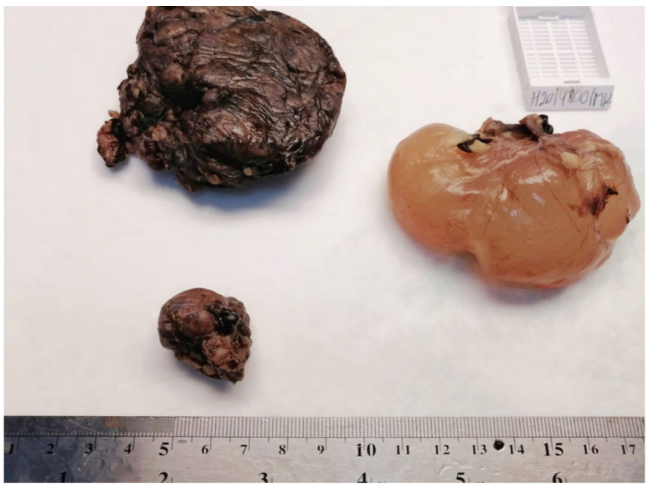

Figure 1. Cystic lesions demonstrating translucent smooth outer surfaces

Microscopic sections showed multicystic lesions with dilated and often tortuous vascular channels filled with serous fluid. They were lined by flattened endothelium without atypia (see Figure 2 and 6), and associated with prominent lymphoid aggregates within the surrounding fibrofatty stroma (see Figure 5). Areas of fibrosis and calcification were also identified (see Figure 3), along with a small amount of attached pancreatic tissue (see Figure 4).

Immunohistochemical sections showed endothelial cells staining positive for CD31, D2-40 (podoplanin), and Factor VIII (see Figures 7, 8 and 9), while negative for pancytokeratin (AE1/AE3). Based on histomorphologic features, immunophenotype and clinical presentation, the findings are compatible with retroperitoneal cystic lymphangiomas.

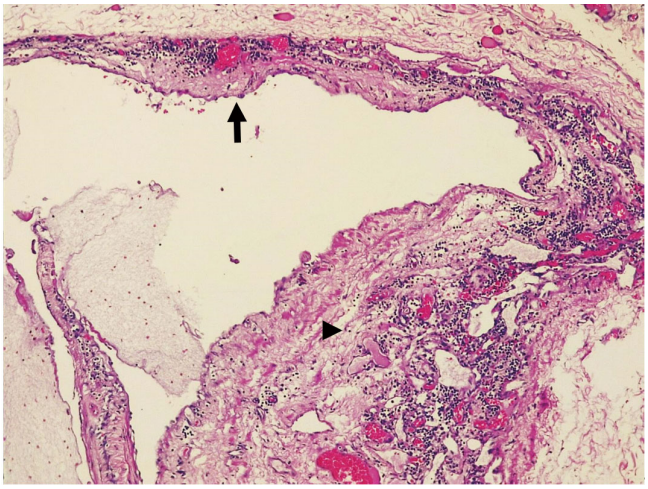

Figure 2. Dilated, tortuous vascular channels filled with serous fluid. Flat endothelial cells without atypia lining these channels (black arrow) Scattered aggregates of small mature lymphocytes are seen within the fibrofatty stroma (arrowheads). (H & E, 10x).

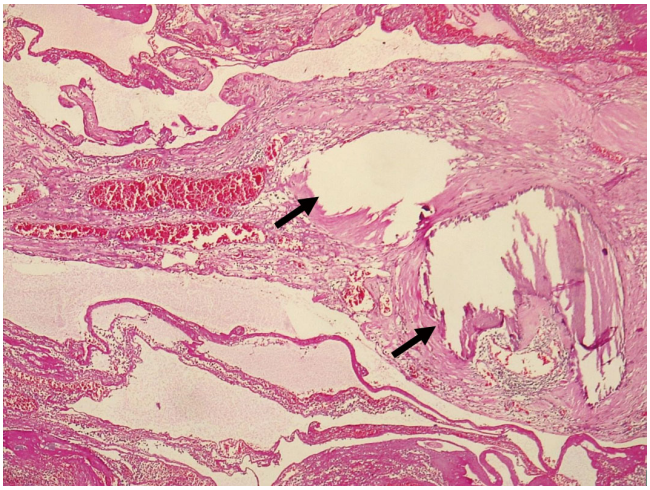

Figure 3. Areas of fibrosis and calcification (black arrow). (H & E, 10x).

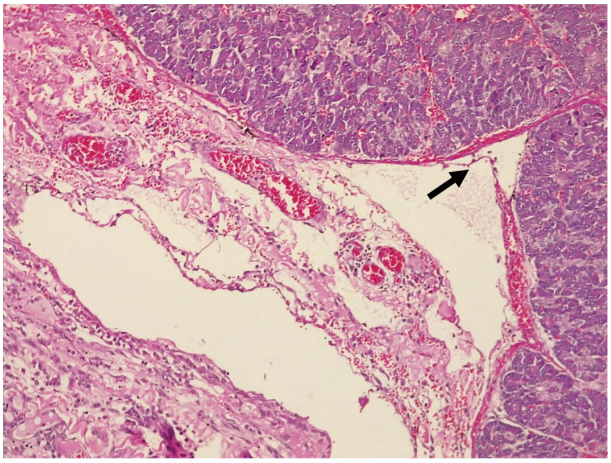

Figure 4. Vascular channels with adjacent pancreatic parenchyma (black arrow). (H & E, 10x).

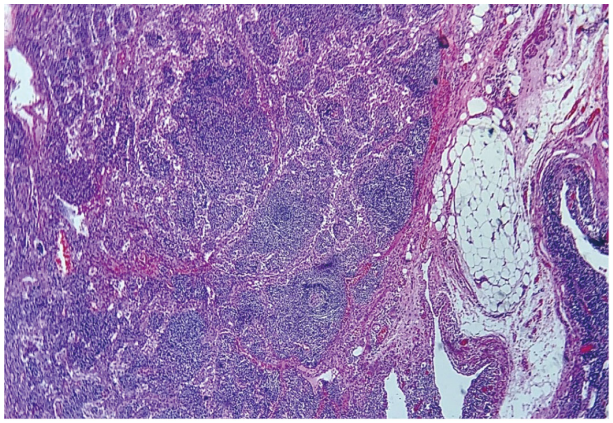

Figure 5. Prominent lymphoid aggregates within the fibrofatty surrounding stroma. (H & E, 4x).

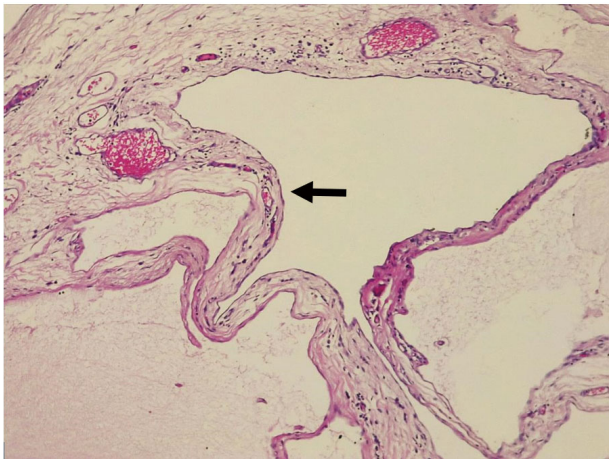

Figure 6. Flat endothelium marked with the black arrow. (H & E, 10x).

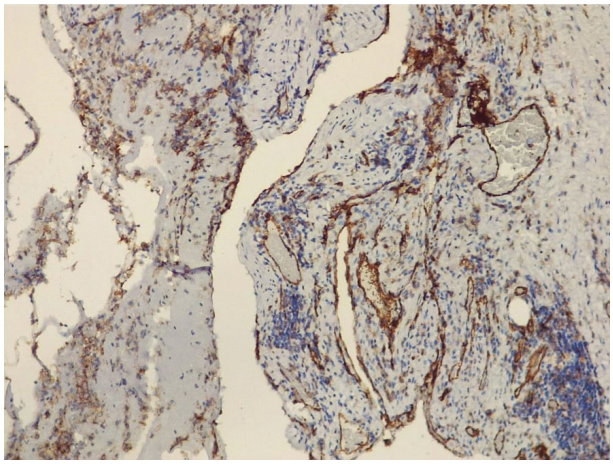

Figure 7. Inmunohistochemical study. CD31 positive in endothelial cells. (10x).

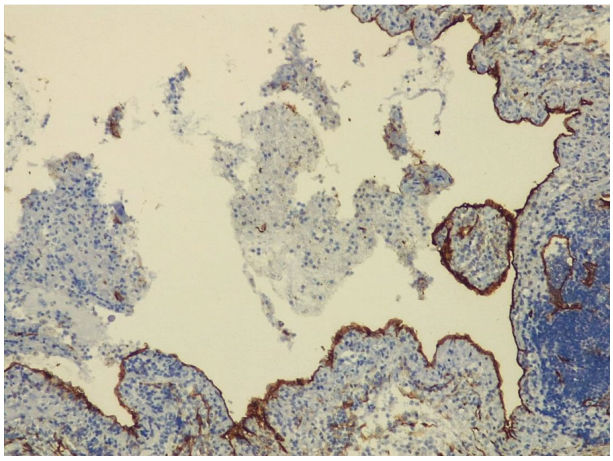

Figure 8. Inmunohistochemical study. D2-40 positive in endothelial cells. (10x).

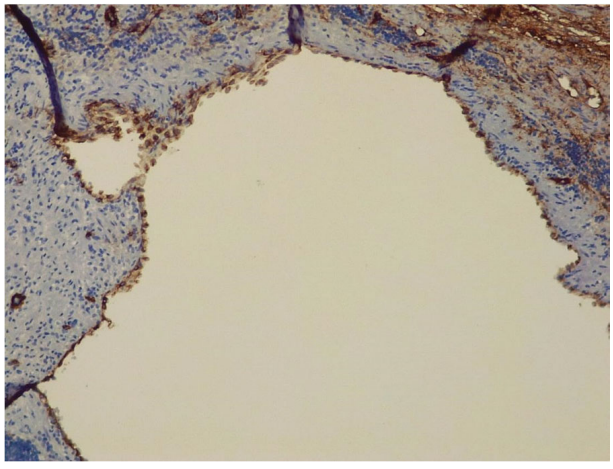

Figure 9. Inmunohistochemical study. Factor VIII positive in vascular channels lined by endothelial cells. (10x).

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3. DISCUSSION

Cystic lymphangiomas are extremely rare benign vascular tumors originating from the lymphatic channels that may involve almost any organ.[1] These are childhood neoplasms with 90% of them being diagnosed before 2 years of age.[2] They are usually seen in the head and neck or axillary region (95%); however, they may rarely be seen in the abdominal cavity of adults, with only 1% occurring in the retroperitoneum.[1,2,5] Adult cystic lymphangiomas are most often seen in women, likely related to oral contraceptive use, pregnancy, or other hormonal influences. These lymphangiomas can reach a large size (approximately 13 cm on average) and cause a palpable mass and abdominal pain.[5]

Our patient presented with abdominal pain, which led to its evaluation through abdominal ultrasound, where multiple cystic masses were evident. Giving the multiple risk factors that endangered the life of the unborn baby, it was decided to intervene and end terminate pregnancy at 32.2 weeks of gestation. The baby was and remained stable and survived. A multidisciplinary approach which include gynecologists, general surgeons, critical care doctors, neonatologists, and pathologic diagnosis was required to address this case. The histological evaluation is the only way to definitely characterize this entity. Few cases of retroperitoneal cystic lymphangiomas have been reported in the literature, especially in pregnant patients.[4,6]

Cystic lymphangiomas are characterized by the presence of multiple dilated cystic spaces lined by flattened endothelium containing abundant lymphoid aggregates and smooth muscle within the cyst wall.[3,5] Histologically, there are 3 distinguishable variants: capillary, cavernous and cystic; with the latter being most common. Papillary endothelial proliferation is a rare finding. Immunohistochemically, the endothelial cells express Factor VIII related antigen, CD31 and CD34, which are endothelial markers without expression of pancytokeratin.[3–5] The differential diagnosis includes dermoid cysts, pseudocysts, and serous cystic neoplasms.[5] The lesions seen in our patient represent a pancreatic cystic lymphangioma, which is a type of retroperitoneal lymphangioma that only represents 0.2% of all pancreatic tumors.

On imaging, they appear as well circumscribed, often multilocular lesions with enhancing capsule and thin septa. The cyst fluid is usually serous, serosanguineous, or chylous. Fine-needle aspiration will show nonspecific features including scattered lymphocytes and histiocytes in a background of amorphous proteinaceous material.[5]

The prognosis of this entity is excellent with total excision; however, recurrence may occur in cases that are incompletely resected.[5,6]

4. CONCLUSION

Cystic lymphangiomas are extremely rare benign neoplasms of lymphatic vessels. There are few cases reported in the literature and even fewer involving pregnant patients. Definitive diagnosis required histologic examination and the prognosis following complete resection is good.

CONFLICTS OF INTEREST DISCLOSURE

There is no conflict of interest.

REFERENCES

[1] Ishibashi Y, Tsujimoto H, Kouzu K, et al. Laparoscopic resection of a huge retroperitoneal cystic lymphangioma after successful reduction of tumor size with a double balloon catheter. Int J Surg Case Rep [Internet]. 2015; 11: 8–10. PMid:25898335. https://doi.org/10.1016/j.ijscr.2015.04.016

[2] Mohammadi A, Ghasemi-rad M, Abassi F. Asymptomatic lymphangioma involving the spleen and mediastinum in adults. Med Ultrason. 2013; 15(2): 154–456. PMid:23702506. https://doi.org/10.11152/mu.2013.2066.152.am1mgr2

[3] Di Marco M, Grassi E, Vecchiarelli S, et al. Retroperitoneal lymphangioma: A report of 2 cases and a review of the literature regarding the differential diagnoses of retroperitoneal cystic masses. Oncol Lett. 2016; 11(5): 3161–6. PMid:27123082. https://doi.org/10.3892/ol.2016.4367

[4] Ozdemir O, Sari ME, Atalay CR, et al. Cystic lymphangioma of the lesser omentum in a pregnant woman: A case report and review of the literature. J Exp Ther Oncol. 2015; 11(2): 155–8. PMid:25667777. https://doi.org/10.1155/2015/583021

[5] Abdelkader A, Hunt B, Hartley CP, et al. Cystic lesions of the pancreas differential diagnosis and cytologic-histologic correlation. Arch Pathol Lab Med. 2020; 144(1): 47–61. PMid:31538798. https://doi.org/10.5858/arpa.2019-0308-RA

[6] Yongo E, Manyala J, Kito K, et al. Diet of Silver Cyprinid, Rastrineobola argentea in Lake Victoria, Kenya. Int J Adv Res. 2016; 4(4): 144–9. https://doi.org/10.21474/IJAR01/113