Adult Splenic Lymphangioma

An Extremely Rare Benign Cystic Lesion of the Spleen

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ABSTRACT

Background: Splenic lymphangioma is a rare, slow-growing benign tumor of the spleen, more often seen in children. However very rarely, they may present in adults also.
When present in adults, it is usually asymptomatic and would be incidentally detected through imaging studies by ultrasound, CT-scan or MRI of abdomen. Infrequently, some patients would present with abdominal pain, distension, nausea, and may have palpable abdominal mass.
Lymphangiomas are benign congenital malformations of the lymphatic vessels that manifest mainly in pediatric patients and infrequently in adults. Usually found in the head and neck areas or axilla. They may involve visceral organs with a lower percentage. The failure of the primordial lymphatics to connect to the main lymphatic vessels or veins. The lymphatic vessels' inability to drain into the veins, the abnormal budding of the lymphatic vessels, and chronic inflammation are some theories to explain the pathogenesis of lymphangiomas.
Accurate radiological diagnosis of splenic lymphangioma is difficult and histopathology with newer immunohistochemistry markers like D2-40 is useful to diagnose splenic lymphangioma.
Surgical removal whether laparoscopic or open is the treatment of choice in large symptomatic splenic lymphangiomas. This is to prevent the life threatening complications like rupture and haemorrhage and to confirm the diagnosis.
Histopathological analysis, the cysts were lined by flat endothelial cells and filled with a pinkish fluid. The cyst walls mainly consisted of hypocellular fibrous tissue with occasional smooth muscle components.
Case summary: A 46-year-old, female obese, body weight 154 kg, with abdominal pain for more than two years, which remained undiagnosed until incidentally discovered on work-up for chronic abdominal pain.
Abdominal ultrasonography revealed splenomegaly with multiloculated cystic mass suggested hydatid cyst of the spleen. Computed tomography with contrast enhancement showed spleen was enlarged in size with evidence of multiple multilocular cysts of variable sizes, largest about 68 x 59 x 54 mm, few were showing peripheral calcifications and showed no enhancement post contrast suggested lymphangioma of spleen.
Surgical splenectomy was performed with eventual resolution of the chronic abdominal pain. Histopathologic examination revealed splenic lymphangioma.

Keywords: Spleen, lymphangioma, Adult, Splenectomy, Immunohistochemistry, Histopathological examination.

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Splenic diseases are very rare, being splenic abscesses, splenic cysts, benign tumors (hemangioma, lymphangioma and others), and malignant tumors (lymphomas, metastases and others)[1]. Primary benign tumors of the spleen are extremely rare and account for less than 0.007% of all tumors identified upon surgery and autopsy[2].

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Splenic lymphangiomas (SL) are benign cystic tumors resulting from congenital malformations of the lymphatic system that appear as single or multiple lesions of the spleen[3-5]. Most lymphangiomas are located in the neck (75%) and armpit (20%), where it is known as cystic hygroma; however, they can appear in other locations (5% of all cases)[6,7], rarely they are found in the spleen[6,8,9].

Splenic lymphangioma is a very rare cystic lesion affecting usually children and

less commonly reported in adult persons[8,10,11].

These lesions are benign and considered by some authors as vascular (lymphatics) malformations rather than true tumors as they are found mainly in children and young patients with some congenital malformative syndrome such as Klippel-Trenaunay syndrome (association of varicose veins, cutaneous capillary malformations and hypertrophy of bone and/or soft tissue)[7,11,12].

The clinical presentation of splenic lymphangiomas is usually related to size of the spleen, but they can be asymptomatic or incidental findings during radiological assessment for other reasons. If they are large, they can cause abdominal pain, loss of appetite, nausea, vomiting and may compress or block adjacent organs.

Clinical and radiological features of splenic lymphangioma are not specific, usually they present as cystic splenic lesions that may correspond to a variety of splenic diseases: congenital epithelial cysts (CEC), neoplastic cysts, parasitic hydatid cysts, traumatic cysts or splenic abscess[11,13-15].

The prognosis is good but there is a very high risk of rupture even after minor trauma, therefore surgery is indicated as definitive treatment.

Surgical removal whether laparoscopic or open is the treatment of choice in large symptomatic splenic lymphangiomas to prevent the life threatening complications like rupture and haemorrhage[16].

Case Report

A 46-year-old female obese, body weight 154 kg, presented to the clinic of general surgery, at Al-Karama general hospital in Baghdad, Iraq, on 25th of November 2023, with a history of more than two years of left upper abdominal pain and this pain was exaggerated in the last three months.

General physical examination was unremarkable and the spleen was not palpable because of the obesity of the patient. Laboratory investigations were within normal limits.

Abdominal ultrasonography revealed a splenomegaly with multiloculated cystic mass (60 x 52 mm), suggested hydatid cyst of the spleen, with normal splenic artery and portal vein.

Computed tomography with contrast enhancement showed enlarged spleen in size with evidence of multiple multilocular cysts of variable sizes, largest about (68 x 59 x 54 mm), some were showing peripheral calcifications, without enhancement post contrast enhancement, (Figure 1).

The patient was scheduled for splenectomy at Al-Zahraa Private Hospital, on 15th of December 2023. Intraoperatively the spleen was found to be enlarged and bosselated as in figure 2.

Splenectomy was performed without any complication and sent for histopathology.

The gross examination showed; huge splenomegaly measured 16.4 cm firm fleshy, greyish-tan to dark-red external surface. The cut section showed irregular thickened fibrotic capsule with central multiloculated microcysts measure 6.9 cm lesion containing clear fluid. No space occupying lesion was seen, with three hailar small nodes vary in size between 0.4-0.6 cm all intact capsule, fleshy cut section.

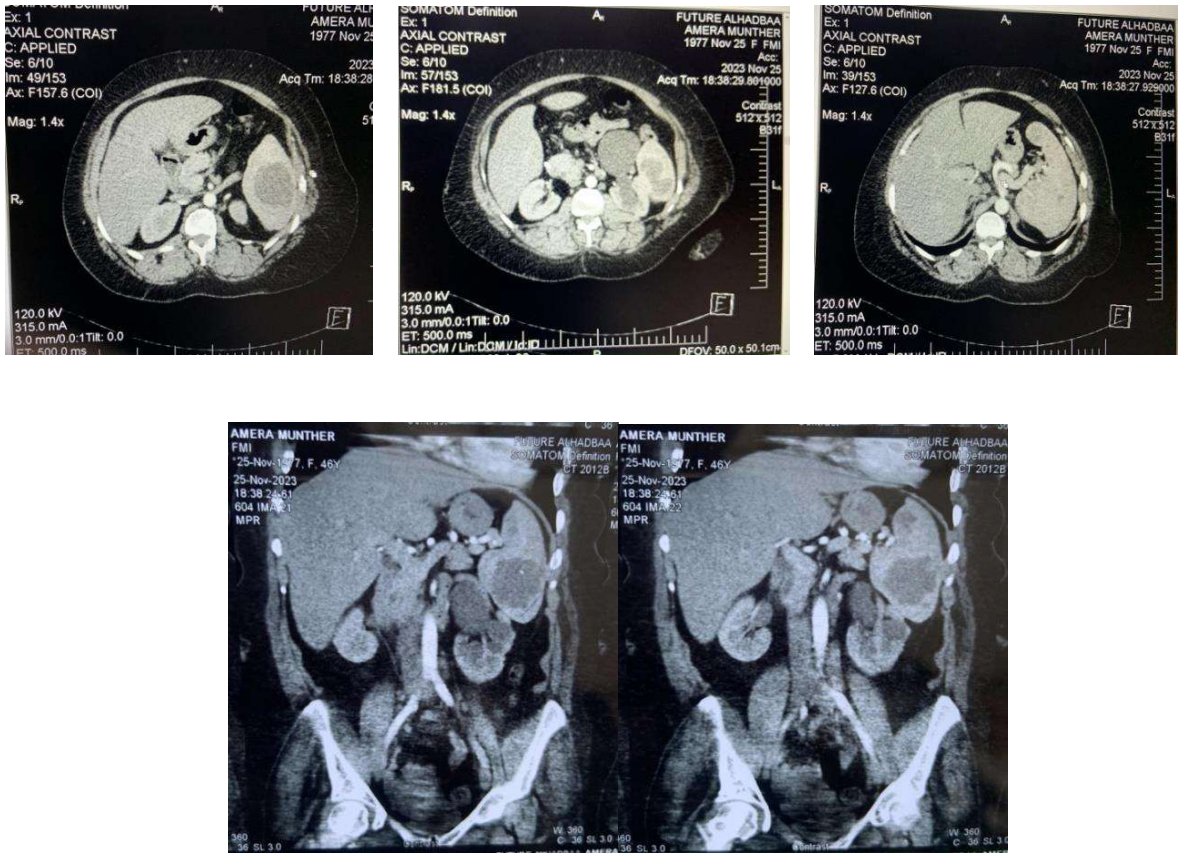

Figure 1: Contrast-enhanced computed tomography showing numerous non-enhancing cystic lesions in an enlarged spleen.

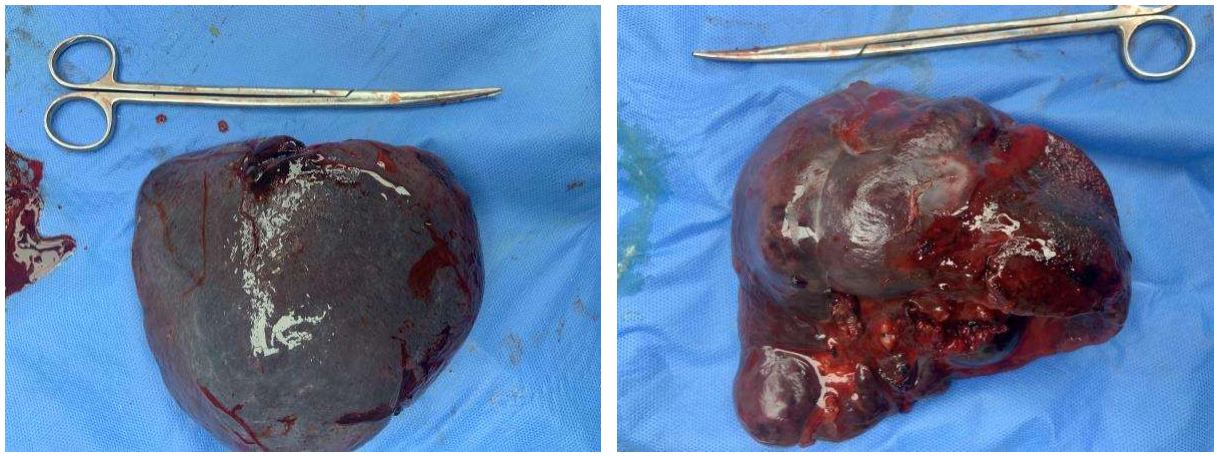

Figure 2: Photographs of splenectomy specimen shows enlarged, nodular and greyish-tan to dark-red external surface with a firm consistency.

Microscopic examination of the lesions showed dilated thin lymphatic vessels containing lymph and intervening stroma showing dense fibrosis. The histopathological features are that of benign lymhangioma of cavernous variant with variable sizes, ectatic lymphatic channels lined by flattened endothelial cells giving positive factor VIII related antigen immunohistochemical endothelial marker.

The surrounding splenic paranchymatous tissue showing

morphological feature of congestive splenomegaly with characteristic thickened fibrous capsule, massive red pulp expansion with marked increase in red cells and hemosiderin laden macrophages, inconspicuous lymphoid white pulp follicle lacking active germinal center, small sclerosiderotic fibrotic nodules (Gamna Gandy bodies) all over the spleen, (Figure 3).

No evidence of TB granuloma or malignancy was seen.

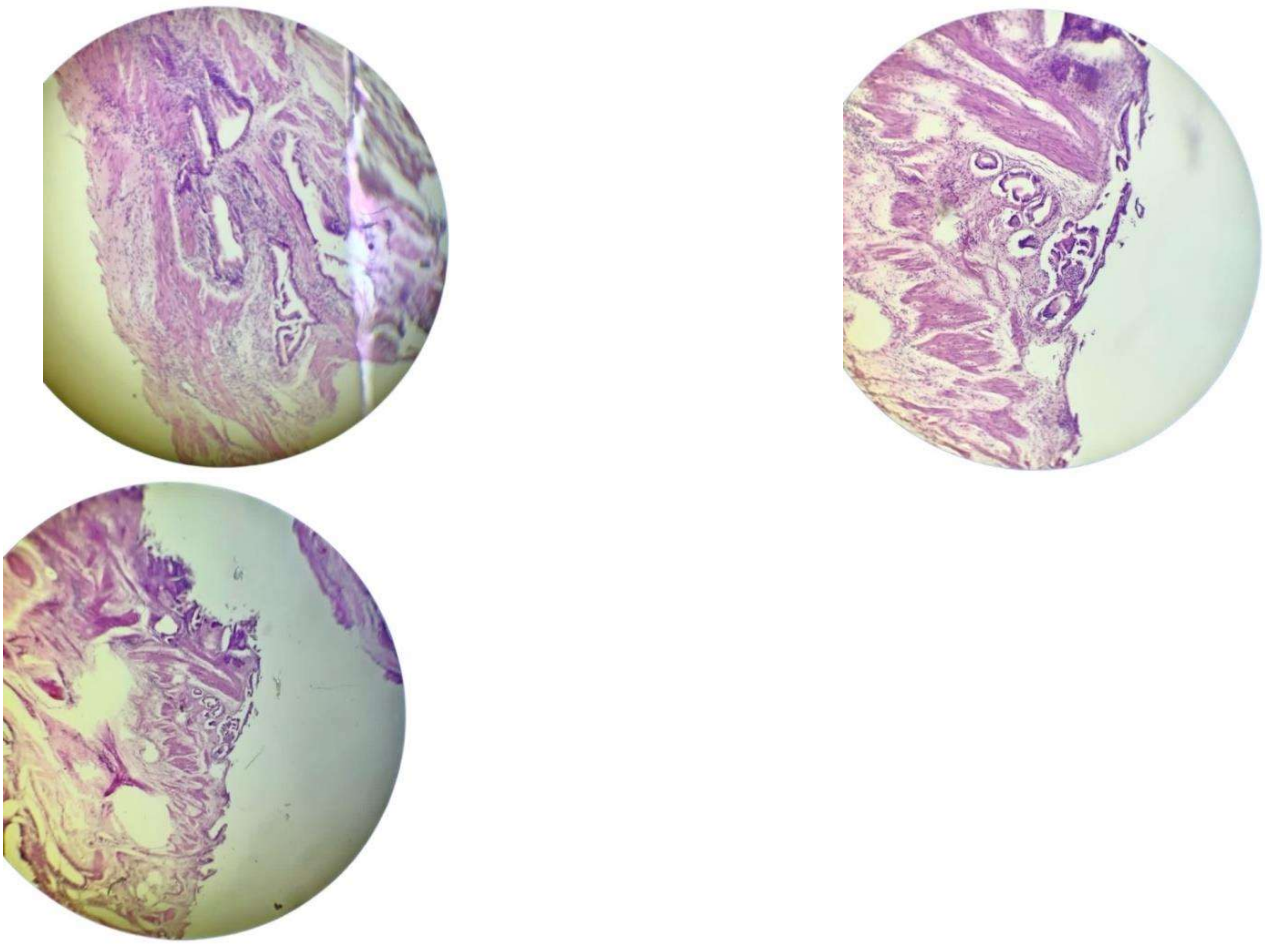

Figure 3: Microscopic examination of the lesions showed dilated thin lymphatic vessels containing lymph and intervening stroma showing dense fibrosis, the lymphatic channels lined by flattened endothelial cells.

Discussion

Splenic lymphangiomas are relatively rare benign tumours that correspond to abnormal dilatation of lymphatic channels that can be either congenital or acquired. It is a slow-growing neoplasm usually seen during childhood and rarely seen beyond 20 years of age[17]. If present in adulthood, it is usually an incidental finding.

The failure of the primordial lymphatics to connect to the main lymphatic vessels or veins, the lymphatic vessels' inability to drain into the veins, the abnormal budding of the lymphatic vessels, and chronic inflammation are some theories to explain the pathogenesis of lymphangiomas.

Most common sites of occurrence of lymphangiomas include the neck,

mediastinum and retroperitoneum[7]. Splenic lymphangiomas are benign cysts that commonly affect children, and are rarely reported in adults[8,10]. In 1885, Frink reported the first lymphangioma in the spleen[18]. Most cases (80-90%) were reported in infants and children less than two years of age[19].

Cystic lesions of spleen include parasitic and non-parasitic cysts. Among parasitic ones, echinococcal disease represents most of the cases[20]. Non-parasitic cysts are primary or true cysts and pseudocysts. Endothelial true cysts include lymphangiomas or hemangiomas[7].

Lymphangiomas can be classified as capillary, cavernous, or cystic[1]. However, the delineation is not universally accepted[21,22].

The clinical manifestations of splenic lymphangiomas are related to the size of lesion. Isolated lesions are asymptomatic and detected incidentally. However, larger lesions may present with abdominal discomfort, loss of appetite or a palpable mass[6,23].

Complications of large lymphangiomas include rupture or infection of the cyst leading to acute abdomen[24]. However rare complications such as consumption coagulopathy, hypersplenism, bleeding, portal hypertension and hypertension due to renal artery compression have also been reported[25].

Diagnosis by radiology is usually not conclusive. In most cases, on CT- scan, splenic lymphangiomas present with thin-walled cystic masses without enhancement or with only slight enhancement of the thin septa. Curvilinear peripheral mural calcifications are suggestive of cystic lymphangiomas but not specific[25].

On T1-weighted magnetic resonance imaging scan, the cystic lesions appear hypointense or hyperintense when filled with hemorrhagic or proteinaceous material[24,25]. On T2-weighted images, they are multiloculated hyperintense corresponding to dilated lymphatic channels[24].

The differential diagnoses of splenic lymphangiomas include other solid and cystic lesions of the spleen, such as hemangioma, chronic infection, lymphoma and metastasis.

Immunohistochemistry (IHC) markers including CD 31, CD 34, factor VIII R Ag, D2-40 and VEGFR-3 may show reactivity in splenic lymphangiomas. Recently, D2-40 has been reported to be a selective marker for the lymphatic endothelium, and can be used for diagnosing splenic lymphangioma accurately from hemangioma also making it a valuable addition for studying benign and malignant vascular disorders in routinely processed tissue specimens[26].

Surgical removal of the spleen in splenic lymphangioma remains the best treatment option, and necessary to relieve symptoms, and prevent complications.

In conclusion: Splenic lymphangioma in adult patients is very rare and should be considered in the differential diagnosis of splenomegaly, and most of the time they are discovered incidentally. Patients may be asymptomatic or present with abdominal pain, nausea, distended abdomen or weight loss.

CT-scan might help to confirming the diagnosis, but the histopathological examination of the resected specimen is the gold standard for definitive diagnosis.

Surgical removal whether laparoscopic or open is the treatment of choice in large symptomatic splenic lymphangiomas to prevent the life threatening complications like rupture and haemorrhage and to confirm the diagnosis.

References

1. Adas G, Karatepe O, Altiok M, Battal M, Bender O, Ozcan D, et al. Diagnostic problems with parasitic and non-parasitic splenic cysts. BMC Surg 2009;29(9):9.
2. Chen LW, Chien RN, Yen CL, Chang LC. Splenic tumour: a clinicopathological study. Int J Clin Pract 2004;58(10):924-7.
3. Chung SH, Park YS, Jo YJ, Kim SH, Jun DW, Son BK, et al. Asymptomatic lymphangioma involving the spleen and retroperitoneum in adults. World J Gastroenterol 2009;15(44):5620-3.

4. de Perrot M, Rostan O, Morel P, C. Abdominal lymphangioma in adults and children. Br J Surg 1998;85(3):395-7.
5. Giovagnoni A, Giorgi C, Goteri G. Tumours of the spleen. Cancer Imaging 2005;25;5(1):73-77.
6. Goh BK, Tan YM, Ong HS, Chui CH, Ooi LL, Chow PK, et al. Intraabdominal and retroperitoneal lymphangiomas in pediatric and adult patients. World J Surg 2005; 29: 837-40.
7. Abbott RM, Levy AD, Aguilera NS, Gorospe L, Thompson WM. From the Archives of the AFIP: Primary vascular neoplasms of the spleen: radiologic-pathologic correlation. Radiographics 2004; 24: 1137-63.
8. Beltrán MA, Barría C, Pujado B, Oliva J, Contreras MA, Wilson CS, et al. Gigantic cystic splenic lymphangioma. Report of one case. Rev Med Chil 2009; 137: 1597-601.
9. Rodríguez-Montes JA, Collantes-Bellido E, Marín-Serrano E, PrietoNieto I, Pérez-Robledo JP4. Splenic lymphangioma. A rare tumour. Presentation of 3 cases and a literature review. Cir Cir 2016; 84: 154-9.
10. M. Vezzoli, E. Ottini, M. Montagne, A. la Fianza, M. Paulli, R. Rosso, et al. Lymphangioma of the spleen in an elderly patient. Haematologica 2000; 85: 314-7.
11. B.K. Goh, Y.M. Tan, H.S. Ong, C.H. Chui, L.L. Ooi, P.K. Chow, et al. Intra-abdominal and retroperitoneal lymphangiomas in pediatric and adult patients. World J Surg 2005; 29: 837-40.
12. D.A. Arber, J.G. Strickler, L.M. Weiss. Splenic mesothelial cysts mimicking lymphangiomas. Am J Surg Pathol 1997; 21: 334-8.
13. B.G. Verghese, S.K. Kashinath, R. Ravi-Kanith. Lymphangioma of the spleen – a rare tumor rarely seen in an adult: a case report and comprehensive literature review. Euroasian J Hepatogastroenterol 2013; 3: 64-69.
14. A. Giovagnoni, C.H. Giorgi, G. Goteri. Tumors of the spleen. Cancer Imaging 2005; 5: 73-77.
15. A.Z. Anadol, M. Oğuz, H. Bayramoğlu, M.N. Edali. Cystic lympahngioma of the spleen mimicking hydatid disease. J Clin Gastroenterol 1998; 26: 309-11.
16. Crema E. Splenic lymphangioma: a rare benign tumor of the spleen treated by laparoscopic surgery. Arq Bras Cir Dig 2012;25(3):178-9.
17. Kaza R., Zara S., Al-Hawary M.M., Francis I.R. Primary and secondary neoplasms of the spleen. Cancer Imaging 2010;10:173-82.
18. Solomou EG, Patriarheas GV, Mpadra FA, Karamouzis MV, Dimopoulos I. Asymptomatic adult cystic lymphangioma of the spleen: Case report and review of the literature. Magn Reson Imaging 2003; 21(1):81-4.
19. Bardeguez A, Chatterjee M, Tepedino M, Sicuranza B. Systemic cystic angiomatosis in pregnancy: A case presentation and review of the literature. Am J Obstet Gynecol 1990; 163(1 Pt 1): 42-5.
20. Lee WS, Kim YH, Chee HK, Lee SA, Kim JD, Kim DC. Cavernous lymphangioma arising in the chest wall 19 years after excision of a cystic hygroma. Korean J Thorac Cardiovasc Surg 2011; 44(5): 380-2.
21. F. Yang and W. X. Chen. Splenic lymphangioma that manifested as a solid-cystic mass: a case report. World Journal of Gastroenterology 2013; 19(5): 781-3.
22. Ogun GO, Oyetunde O, Akang EE. Cavernous lymphangioma of the breast. World J Surg Oncol 2007;5:69.
23. Komatsuda T, Ishida H, Konno K, Hamashima Y, Naganuma Y, Sato M et al. Splenic Lymphangioma: US and CT diagnosis and clinical manifestations. Abdom Imaging 1999; 24 (4):414-7.
24. Moir C, Guttman F, Jequier S, Sonnino R, Youssef S. Splenic cysts: aspiration, sclerosis, or resection. J Pediatr Surg 1989; 24(7): 646-8.
25. Dietz WH, Stuart MJ. Splenic consumptive coagulopathy in a patient with disseminated lymphangiomatosis. J Pediatr 1977; 90 (3):421-3.
26. Takayama A, Nakashima O, Kobayashi K, Kojiro M. Splenic lymphangioma with papillary endothelial proliferation: A case report and review of the literature. Pathol Int 2003; 53(7): 483-8.

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