

## Case Report

# Splenic Lymphangioma Mimicking Lymphomatous Involvement: A Case Report with Review of the Literature

**Neda Soleimani** <sup>1,2</sup> **Fatemeh Pouraminaee** <sup>3</sup> **Mohammad Hossein Anbardar** <sup>1,2</sup>  
**Ali Bahador** <sup>2</sup> **Benyamin Rahimi** <sup>1</sup> **Sahand Mohammadzadeh** <sup>1,2</sup>  
**Fatemeh Aghakhaninejad** <sup>1</sup> **Mohammad Farahmand** <sup>2</sup> and **Mahsa Hasani** <sup>1</sup>

<sup>1</sup>Department of Pathology, Shiraz Medical School, Shiraz University of Medical Sciences, Shiraz, Iran

<sup>2</sup>Shiraz Transplant Center, Abu Ali Sina Hospital, Shiraz University of Medical Sciences, Shiraz, Iran

<sup>3</sup>Pathology Research Center, Shiraz University of Medical Sciences, Shiraz, Iran

Correspondence should be addressed to Neda Soleimani; [soleimani\\_n@sums.ac.ir](mailto:soleimani_n@sums.ac.ir)

Received 5 January 2023; Revised 8 June 2023; Accepted 14 June 2023; Published 20 June 2023

Academic Editor: Simon Ching Shun Kao

Copyright © 2023 Neda Soleimani et al. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Lymphangioma is a benign malformation of lymphatic vessels usually found in the head and neck areas or axilla. They may involve visceral organs with a lower percentage. Splenic lymphangioma is a rare tumor. This disease is often seen in children but may be diagnosed incidentally in adults. Most patients are asymptomatic, but in large and multifocal lesions, the patient may have some nonspecific symptoms such as abdominal pain, abdominal distention, nausea, vomiting, and loss of appetite. Physical examination may show no specific findings or detect palpable masses. The preoperative diagnosis of splenic lymphangioma is challenging. Histopathological evaluation and sometimes immunohistochemistry tests can result in a definitive diagnosis. In this study, we present an 18-year-old man, with Burkitt's lymphoma who underwent laparotomy and total splenectomy as a result of cystic lesions discovered accidentally during imaging with the final diagnosis of splenic lymphangioma after histopathological evaluation.

## 1. Introduction

Lymphangioma is a benign neoplasm of lymphatic vessels, often affecting children's head and neck areas. It is uncommon to find lymphangiomas in abdominal organs such as the kidney, adrenal, small bowel mesentery, liver, and spleen. Splenic lymphangioma is very rare and is often found incidentally. Nevertheless, some patients may experience nonspecific signs and symptoms such as abdominal pain and distention. Preoperative diagnosis would be difficult, and overdiagnosis might happen because radiological findings are also not specific. The ultimate diagnosis is usually made by histopathological examination [1–7]. Herein, we report a case of splenic lymphangioma in an adult patient who had a previous case of Burkitt's lymphoma.

## 2. Case Report

An 18-year-old man was transferred to our center to be evaluated for splenic lesions, which were found incidentally during a routine annual follow-up for Burkitt's lymphoma. He had no history of abdominal pain, fever, or weight loss. He has been a case of Burkitt's lymphoma of the stomach since seven years ago and has received chemotherapy for one year. There was no sign of a disease recurrence, and all earlier ultrasonographic evaluations were unremarkable until the last follow-up, which revealed splenic lesions. Other past medical and family histories were not significant. On physical examination, blood pressure was 110/70 mmHg with a normal pulse rate and temperature, 68 beats per minute and 37.1°C, respectively. The abdomen was soft, and there was no organomegaly. Complete blood count (CBC) (white blood

cell (WBC):  $4.9 \times 10^3/\mu\text{l}$ , hemoglobin: 14.2 g/dl, and platelet count:  $211 \times 10^3/\mu\text{l}$ ) and liver enzymes, including aspartate aminotransferase (AST), alanine aminotransferase (ALT), and alkaline phosphatase (ALP), were within normal ranges. Abdominopelvic ultrasonography revealed a hyperechoic mass measuring 24 mm and a cystic lesion measuring 9 mm in the spleen parenchyma. A spiral CT scan of the abdomen with contrast showed a few near together cystic lesions up to 18 mm in the anterosuperior aspect and a hypoattenuated subcapsular lesion measuring 24 mm in the posterior aspect of the spleen with faint enhancement after administration of contrast, suggestive of lymphomatous infiltration (Figure 1). The patient underwent laparotomy. The splenic lesions were multiple at the time of the operation, so a diagnostic splenectomy was performed for him. Gross sectioning of the spleen showed two multiloculated cystic lesions, the largest measuring  $1.5 \times 1 \times 1$  cm and containing some brownish fluid (Figure 2). Histological examination showed multiple large irregular cystic spaces lined by endothelial cells with smooth pinkish fluid in the lumen and lymphoid aggregation consisting of lymphangiomas (Figures 3(a) and 3(b)). Two months later, no clinical or radiologic deficits were noted in a follow-up visit. We obtained the patient's informed consent for publishing the case report and images. Our institutional approval was not required to publish the case details.

### 3. Discussion

Splenic lymphangioma is a benign cystic neoplasm initially described by Frink in 1885, accounting for <0.007% of all tumors. It typically affects pediatric patients but is rarely found in adults [6, 8].

Sequestration of lymph tissue, aberrant lymph vessels budding, lack of integration with the venous system, and obstruction of lymph vessels are some theories to explain the pathogenesis of lymphangiomas.

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 a few of the hypotheses that have been suggested for the etiopathogenesis of lymphangiomas. Vascular endothelial growth factor-C (VEGF-C) and vascular endothelial growth factor-R3 are also elevated. However, in adult patients, as in our case, only a tiny defect often exists in the lymphatic system, which can be compensated under healthy conditions. Pathologic conditions associated with an increased lymphatic volume may then result in the development of lymphangioma. There is speculation that trauma and infectious diseases are the catalysts for this development [6, 9, 10]. Similarly, the fact that our patient developed a new splenic lesion that was not present in previous radiologic studies suggests that prior surgery and chemotherapy may have served as a trigger.

Clinical presentations depend on the tumor size. Most patients are asymptomatic, and the tumor is found incidentally. Large and multifocal lesions can, however, cause nonspecific symptoms such as abdominal pain, abdominal distention, nausea, vomiting, and loss of appetite, which are primarily brought on by the compression of nearby organs.

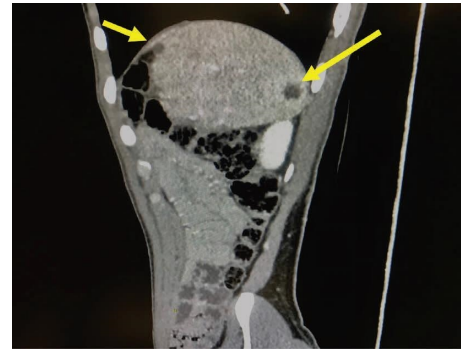


FIGURE 1: Abdominal spiral CT scan with contrast shows a few near together cystic lesions with faint enhancement after administration of contrast (arrows).



FIGURE 2: Cut section of the spleen shows two well-defined multiloculated cystic lesions (arrows).

Post-traumatic and spontaneous rupture of splenic lymphangiomas has also been reported. Moreover, it can lead to coagulopathy, hemorrhage, and portal hypertension. Physical examination is unremarkable or may detect a palpable mass [1, 11–13].

Radiological findings are generally not specific. Splenic lymphangiomas in ultrasonography may appear as anechoic, hypoechoic, or hyperechoic lesions depending on whether the lymphatic fluid is infected, hemorrhagic, or hyperlipidemic. They have different diameters, with or without internal septation. Calcification is a nonspecific finding and may also be observed in splenic hydatid cysts. Like in our case, computed tomography (CT) scan reveals single or multiple well-defined thin-walled lesions that are usually hypodense, homogenous, and without significant contrast enhancement [2, 12, 14]. The lesions are often well-vascularized in angiography.

Magnetic resonance imaging (MRI) shows hypointense cystic lesions in T1-weighted images, yet on T2-weighted images, they look hyperintense when filled with hemorrhagic or proteinaceous material. There have been some reported instances of lymphangioma without F-18-fluorodeoxyglucose (FDG) uptake on positron emission tomography (PET) scans. However, they may exhibit slightly enhanced F-18 FDG uptake as a result of lymphocytes and fluid joining the dilated lymphatic vessels [1, 6, 15–19].

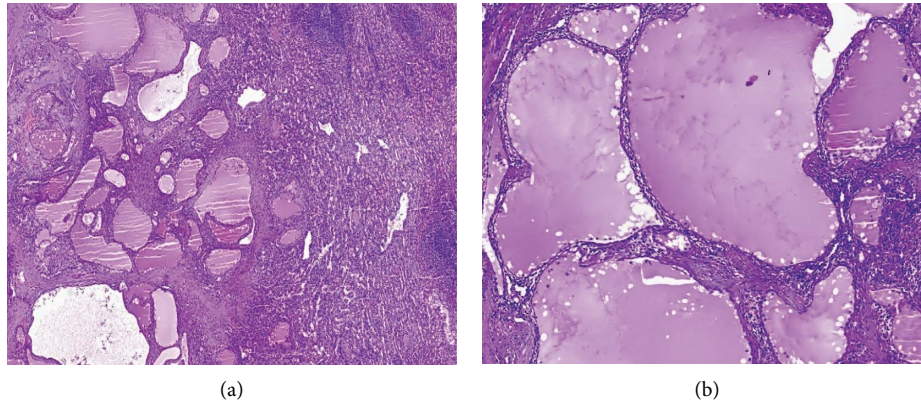


FIGURE 3: Microscopic assessment shows vascular spaces containing pinkish fluid (a) (H&E  $\times 40$ ). The spaces are lined by endothelial cells (b) (H&E  $\times 200$ ).

Gross appearance shows single or multiple cystic lesions in various sizes with a thick fibrous wall filled with clear fluid. Multiple lesions may have a honeycomb look. Histopathological evaluation reveals cystic lesions lined by endothelial cells and filled with an amorphous eosinophilic proteinaceous substance [2, 20, 21]. Moreover, lymphangiomas are divided into three categories based on the size of the dilated lymphatic channels: (1) capillary (supermicrocystic), (2) cavernous (microcystic), and (3) cystic (macrocytic) [6].

Immunohistochemistry can be used when there is a doubt about the ability to differentiate cystic lesions of splenic lymphangioma from other cystic lesions such as mesothelial cysts, epidermoid cysts, and parasite cysts. The cells lining the cystic walls in splenic lymphangioma stain positive for CD34, CD31, factor VIII, vascular endothelial growth factor 3, and podoplanin [22–24].

Total splenectomy is the recommended treatment choice after the diagnosis is confirmed to reduce the risk of consequences such as infection, bleeding, growth, and expansion of the lesions and spleen rupture. Splenic lymphangioma has a favorable outcome after total splenectomy, and the possibility of malignancy is scarce [1, 3, 5].

Our patient's history of Burkitt's lymphoma resulted in a misdiagnosis of lymphomatous involvement in a CT scan study. However, if used, MRI and PET scans could aid with a more precise diagnosis and prevent misdiagnosis. In addition, since the lesions were located superficially in the spleen, a more accurate preoperative diagnosis might result in cyst excision rather than an unnecessary total splenectomy.

#### 4. Conclusion

Splenic lymphangioma is an extremely rare tumor. Although this benign entity frequently affects children, it should be taken into consideration as a differential diagnosis in adults with acquired cystic lesions, especially in those who have already experienced intra-abdominal stress (surgery, trauma, infection, and so on).

#### Abbreviations

CBC: Complete blood count  
 WBC: White blood cell  
 AST: Aspartate aminotransferase  
 ALT: Alanine aminotransferase  
 ALP: Alkaline phosphatase  
 CT: Computed tomography  
 MRI: Magnetic resonance imaging  
 FDG: Fluorodeoxyglucose  
 PET: Positron emission tomography.

#### Data Availability

All data generated or analyzed during this study are included in this published article.

#### Consent

A written informed consent was obtained by the patient for publishing the case report and the publication of the accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

#### Conflicts of Interest

The authors declare that they have no conflicts of interest.

#### Authors' Contributions

SN, RB, and PF developed the concept of the study and the study design and wrote the draft manuscript. RB extracted the history. BA performed the operation. SN, PF, AF, MS, AF, and SM with the input of all authors interpreted the patient data. SN and AMH reviewed the pathologic slides and analyzed all data. All authors have read and approved the final manuscript.

#### References

- [1] I. Ioannidis and A. G. Kahn, "Splenic lymphangioma," *Archives of Pathology & Laboratory Medicine*, vol. 139, no. 2, pp. 278–282, 2015.

- [2] M. Bisceglia, A. Chiaramonte, W. Lauriola, C. A. Galliani, P. Musto, and G. Colacchio, "Selected case from the Arkadi M. Rywlin international pathology slide series: lymphangiomatosis of the spleen associated with ipsilateral abdominopelvic and lower extremity venolymphatic malformations: a case report and review of the literature," *Advances in Anatomic Pathology*, vol. 21, no. 4, pp. 291–299, 2014.
- [3] E. Crema, R. M. Etchebehere, M. N. Gonzaga, R. S. Lima, P. A. Bertulucci, and A. A. Silva, "Linfangioma esplênico: um raro tumor benigno do baço tratado por cirurgia laparoscópica," *ABCD. Arquivos Brasileiros de Cirurgia Digestiva (São Paulo)*, vol. 25, no. 3, pp. 178–179, 2012.
- [4] S. Y. Kim, H. J. Kwon, H. W. Park, S. Y. Lee, B. H. Son, and M. S. Kim, "Multiple cystic lymphangiomas of the spleen: radiologic and histopathologic findings," *Journal of Medical Ultrasonics*, vol. 42, no. 3, pp. 409–412, 2001.
- [5] R. Patti, E. Iannitto, and G. Di Vita, "Splenic lymphangiomatosis showing rapid growth during lactation: a case report," *World Journal of Gastroenterology*, vol. 16, no. 9, pp. 1155–1157, 2010.
- [6] A. Perez, M. E. C. Perez, A. C. Yuga, and B. A. G. Viray, "Splenic lymphangioma in adulthood: a case report," *International Journal of Surgery Case Reports*, vol. 67, pp. 250–253, 2020.
- [7] L. W. Chen, R. N. Chien, C. L. Yen, and L. C. Chang, "Splenic tumour: a clinicopathological study," *International Journal of Clinical Practice*, vol. 58, no. 10, pp. 924–927, 2004.
- [8] F. Yang and W. X. Chen, "Splenic lymphangioma that manifested as a solid-cystic mass: a case report," *World Journal of Gastroenterology*, vol. 19, no. 5, pp. 781–783, 2013.
- [9] S. Hasan, S. A. Ahmad, M. Kaur, R. Panigrahi, and S. Panda, "Lymphangioma of the lower lip-A diagnostic dilemma: report of a rare case with a brief literature review," *Case Reports in Dentistry*, vol. 2022, Article ID 7890338, 8 pages, 2022.
- [10] S. Wiegand, B. Eivazi, P. J. Barth et al., "Pathogenesis of lymphangiomas," *Virchows Archiv*, vol. 453, no. 1, pp. 1–8, 2008.
- [11] G. Evola, G. Mazzone, A. Corsaro, G. Brancato, F. R. Evola, and G. Basile, "Hemorrhagic shock from post-traumatic rupture of microcystic splenic lymphangioma: a case report and review of the literature," *International Journal of Surgery Case Reports*, vol. 75, pp. 376–379, 2020.
- [12] R. M. Abbott, A. D. Levy, N. S. Aguilera, L. Gorospe, and W. M. Thompson, "From the archives of the AFIP: primary vascular neoplasms of the spleen: radiologic-pathologic correlation," *RadioGraphics*, vol. 24, no. 4, pp. 1137–1163, 2004.
- [13] S. H. Chung, Y. S. Park, Y. J. Jo et al., "Asymptomatic lymphangioma involving the spleen and retroperitoneum in adults," *World Journal of Gastroenterology*, vol. 15, no. 44, pp. 5620–5623, 2009.
- [14] D. Diarra, Y. Mekouar, A. Salihou et al., "Cystic lymphangioma of the upper limb: clinical case and literature review," *BJR case reports*, vol. 8, no. 5, Article ID 20210206, 2022.
- [15] M. Eghtedari, J. Sicklick, Y. Kono, M. R. Peterson, and C. S. Santillan, "Unusual imaging profile of a solitary splenic lymphangioma," *Acta Radiologica Short Reports*, vol. 1, no. 8, pp. 1–5, 2012.
- [16] M. H. Anbardar, N. Soleimani, A. Aminzadeh Vahedi, and S. A. Malek-Hosseini, "><>," *International Medical Case Reports Journal*, vol. 12, pp. 297–301, 2019.
- [17] M. Bezzi, A. Spinelli, M. Pierleoni, and G. Andreoli, "Cystic lymphangioma of the spleen: US-CT-MRI correlation," *European Radiology*, vol. 11, no. 7, pp. 1187–1190, 2001.
- [18] M. M. Sun and J. Shen, "Positron emission tomography/computed tomography findings of multiple cystic lymphangiomas in an adult: a case report," *World Journal of Clinical Cases*, vol. 8, no. 10, pp. 1973–1978, 2020.
- [19] T. Ji and A. Kuang, "18F-FDG PET/CT findings in a splenic lymphangioma," *Clinical Nuclear Medicine*, vol. 40, no. 7, pp. e375–e377, 2015.
- [20] S. Duvvada, D. Senapati, S. R. Challa, and T. Kalluri, "Cystic lymphangioma of spleen in adults," *BMJ Case Reports*, vol. 2017, Article ID bcr2016216267, 2017.
- [21] W. C. Chang, C. H. Liou, H. W. Kao, C. C. Hsu, C. Y. Chen, and C. Y. Yu, "Solitary lymphangioma of the spleen: dynamic MR findings with pathological correlation," *British Journal of Radiology*, vol. 80, no. 949, pp. e4–e6, 2007.
- [22] L. V. Debelenko, A. R. Perez-Atayde, J. B. Mulliken, M. G. Liang, T. H. Archibald, and H. P. Kozakewich, "D2-40 immunohistochemical analysis of pediatric vascular tumors reveals positivity in kaposiform hemangioendothelioma," *Modern Pathology*, vol. 18, no. 11, pp. 1454–1460, 2005.
- [23] D. A. Arber, J. G. Strickler, and L. M. Weiss, "Splenic mesothelial cysts mimicking lymphangiomas," *The American Journal of Surgical Pathology*, vol. 21, no. 3, pp. 334–338, 1997.
- [24] D. Sethi, M. Garg, S. Kataria, and S. K. Mathur, "Epidermoid cyst of spleen mimicking splenic lymphangioma," *Advanced Biomedical Research*, vol. 2, no. 1, p. 49, 2013.