JK SCIENCE pISSN: 0972-1177

CASE REPORT

Isolated Splenic Cystic Lymphangioma in an Adult: A Diagnostic Dilemma

Rashmi Ranjan Sahoo, Ketan Prabhunath Gupta, Nalini Kumar Naik, Deepankar Majhi

Abstract

Lymphangiomas are rare congenital malformations of the lymphatic system, seen in children and infrequently in adults. They most commonly involve the neck (75%) and axilla (20%), with abdominal involvement accounting for less than 5%. Spleniclymphangiomas are exceptionally rare, with fewer cases reported in the literature. They are often asymptomatic and incidentally detected, posing diagnostic challenges due to their resemblance to other cystic splenic lesions such as hydatid disease. We report a rare case of isolated spleniclymphangioma, highlighting its diagnostic ambiguity and the importance of timely intervention once diagnosed.

Key Words

Cystic, Lymphangioma, Spleen

Introduction

Lymphangiomas are uncommon benign neoplasms resulting from congenital malformations of the lymphatic system. While 95% of these lesions occur in the neck and axilla during childhood, intra-abdominal involvement is exceedingly rare, representing less than 5% of cases.[1-3]Among these, isolated splenic cystic lymphangioma in adults is a singular entity. It accounts for 0.007% of all tumors.[4] The first case involving the spleen was reported in 1885 by Frink.[5] Between 1939 and 2017, only 209 cases of spleniclymphangiomas were reported in the literature.[6] In most patients, the lymphangiomatous process involves additional sites in a diffuse or multifocal fashion such as the liver, mediastinum, and lung, the so-called lymphangiomatosis syndrome.[7] Only 20 cases of isolated splenic lymphangiomas were reported between 1990 and 2024.[8] This case highlights the diagnostic uncertainty surrounding splenic cystic lesions and emphasizes the necessity of histopathological and immunohistochemical (IHC) confirmation following surgical intervention.

Case Report

A 43-year-old married woman came to outpatient department of General Surgery, in a tertiary care centre with complaints of intermittent abdominal pain, intermittent vomiting, dyspepsia, and some loss of appetite for 2 years. The patient had a history of irregular menses. The patient was well-nourished and had a normal build. An abdominal ultrasound was performed for irregular menses and epigastric pain, which revealed a multicystic lesion in the spleen (hydatid cyst). A contrast enhanced computed tomography(CECT) scan was subsequentlyadvised for further evaluation. CECT abdomen and pelvis showed a well-defined, non-enhancing, multiloculated cystic lesion in the lower pole of the spleen (nature uncertain). The spleen was normal in shape, slightly enlargedin size. The lesion measured 65x55x48 cm (APxMLxCC) in the lower

Department of General Surgery, Institute of Medical Sciences and SUM Hospital, Siksha O Anusandhan Deemed to be University, Bhubaneswar, Odisha, India

Correspondence to: Dr. Ketan Gupta, Department of General Surgery, Institute of Medical Sciences and SUM Hospital, Siksha O Anusandhan Deemed to be University, Bhubaneswar, Odisha, India.

Manuscript Received: 08.01.2026; **Revision Accepted**: 07.04.2026;
Published Online First: 10th July, 2026.
Open Access at: https://journal.jkscience.org

Cite this article as: Sahoo RR, Gupta KP, Naik NK, Majhi D. Isolated Splenic Cystic Lymphangioma in an Adult: A Diagnostic Dilemma. JK Science 2026;28(3):199-201.

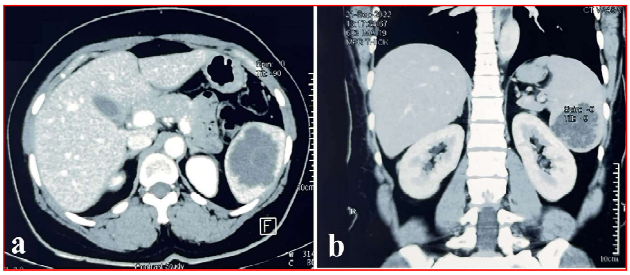

Figure 1:
(a) Axial section of CECT showing cystic lesion in spleen.
(b) Coronal section of CECT.

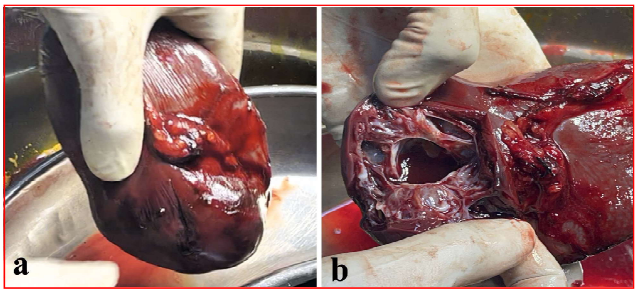

Figure 2:
(a) Lower pole of spleen showing lesion.
(b) Cystic spaces with gelatinous material.

pole, and the splenic vein was normal in caliber. There was no evidence of any lymphadenopathysuggestinghydatid disease of spleen (Figure 1a & 1b). Due to diagnostic uncertainty, a splenectomy was planned. The general examination findings were unremarkable. On palpation, the abdomen was soft and nontender. The spleen's inferior edge was palpable, extending to a point of 2 centimeters below the left costal margin.All routine investigations were normal. Pre-ophaemoglobin level was 10.5 g/dL. Preoperative vaccination was done against encapsulated organisms. Due to the likely diagnosis of a hydatid cyst of the spleen, an open splenectomy was planned. It revealed a multicystic lesion on the lower pole filled with gelatinous material. There was no evidence of daughter cysts, hydatid sand, germinal layer, or hooklets. On histopathological examination, the spleen measured 12x8x4 cm, weighing around 400 gm. The outer surface was smooth, and the glistening capsule was intact. On serial sectioning, fleshy multiple cystic areas were filled with greenish and yellowish gelatinous material, with the largest cyst measuring 1 cm in the greatest dimension. Cystic areas were present along with normal fleshy splenic parenchyma (Figure 2a & 2b). Microscopically, sections showed the red pulp and white pulp of the spleen,

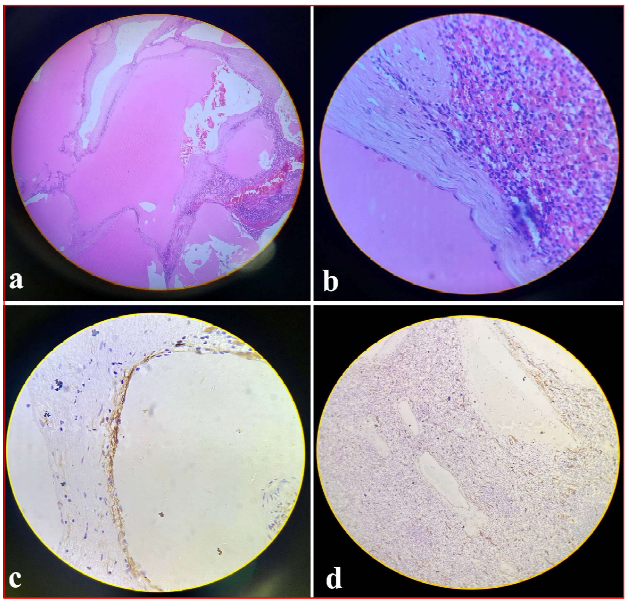

Figure 3:
(a) Cystic spaces lined by flattened epithelium and containingeosinophilicproteinaceousmaterial. [100X H&E]
(b) Fibrous septa and splenicparenchyma. [400X H&E]
(c) IHC staining for CD34 staining the continuous endothelial lining of the dilated vascular spaces.[400X IHC CD34]
(d) Low power image [100X] of the endothelium stained with IHC CD34

along with a lesional componentcomprised of multiple cystic spaces of varying sizes. These cystic spaces were lined by flattened endothelial cells and filled with eosinophilic proteinaceous fluid. No evidence of any organism or malignancy was seen in the sections (Figure 3a & 3b). The findings were consistent with cystic lymphangioma of the spleen and confirmed with IHC staining with CD34, which came out to be positive (Figure 3c & 3d). The postoperative period was marked bypost-splenectomyleukocytosis count (TLC count 24,000) on postoperative day (POD)1. The TLC count returned to normal values by POD 3. The patient was discharged on POD 5. The patient had follow-up appointments after 2 weeks and 2 months, respectively, with an abdominal ultrasound, which showed no abnormalities. The patient made a full recovery.

Discussion

Lymphangiomas are benign neoplasmsthat result from malformation of the lymphatic system. Other hypotheses

suggest that inflammation of the lymphatic system can cause obstruction and subsequent formation of lymphangiomas. Solitary spleniclymphangioma is typically asymptomatic. However, large cystic lesions may cause splenomegaly and manifest as left upper quadrant LUQ pain, intermittent epigastric pain, distension, loss of appetite, vomiting, palpable mass, chronic back pain, and fever.[8] In cystic lymphangiomas, the clearly outlined cysts typically observed in the subcapsular regions exhibit a hypodense appearance, featuring sporadic enhancing septa and occasional calcification around the periphery. On magnetic resonance imaging (MRI), they manifest as well-defined, multilocular cystic formations with delicate partitions that display uniform hypo intensity on T1-weighted images. On T2-weighted images, they appear hyperintense when filled with hemorrhagic or proteinaceous material. Due to their multiloculated cystic appearance on imaging, they often mimic more common pathologies such as hydatid disease, hemangiomas, or pseudocysts. Cystic lesions of the spleen have radiological similarities and are therefore difficult to diagnose. Microscopically, these cysts are made up of variably sized, thin-walled vascular channels lined by a single layer of endothelial cells and filled with eosinophilic, proteinaceous material. IHC staining includes FVIII-Rag, CD31, CD34 and D2-40 (a selective marker for lymphatic endothelium) for the identification of endothelial cell lining.[5] The distinctive spleen appearance, often likened to that of "Swiss cheese," is often considered pathognomonic.FNAC (Fine-Needle Aspiration Cytology) is contraindicated due to thesubstantial risk of bleeding and insufficient tissue sampling for making a diagnosis.[10]Postoperative histological and immunohistochemical techniques can be used to confirm the diagnosis. Splenectomy is the treatment of choice, with laparoscopic methods being preferred. Early intervention is necessary to prevent complications such as rupture with peritonitis, invasive haemorrhage, infection, and abscess formation. Large spleens are associated with bleeding and consumptive coagulopathy, hypersplenism, portal hypertension, and diaphragmatic immobility with atelectasis. The malignant transformation rate is low, and surgery is associated with a good prognosis and low recurrence.

Conclusion

Cystic lymphangiomas necessitate a comprehensive and multidisciplinary diagnostic approach to enable timely intervention and distinguish them from other cystic conditions. The integration of molecular and histopathological diagnostic modalities assumes paramount importance in this case, serving the crucial purpose of ruling out alternative pathologies characterized by adverse prognoses, including malignancy, its rarity, cystic lymphangioma warrants profound consideration as a significant differential diagnosis for cystic splenic lesions, owing to the inherent diagnostic ambiguities involved and its pivotal role as an imperative indication for surgical intervention aimed at averting future complications.

Financial Support and Sponsorship : Nil
Conflict of Interest: Nil

References

1. Al-Janabi MH, Abodest R, Alshabab MA, Kannan S, Issa R. Splenic cystic lymphangioma in adults: A rare case report from Syria. Annals of Medicine and Surgery. 2022 Aug 1;80.
2. Ismail IB, Yaakoubi J, Helal I, Tounsi MK, Sghaier M, Zoghlami A. Symptomatic diffuse cystic lymphangioma of the spleen in an adult: A case report. International Journal of Surgery Case Reports. 2025 Jan 1;126:110669.
3. Shi R, Yang P, Guo Y, Tang Y, Luo H, Qin C, et al. Laparoscopic anatomical partial splenectomy for extremely rare isolated splenic lymphangioma in an adult: a case report and literature review. Frontiers in Oncology. 2024 Sep 4;14:1460566.
4. Alessa H, Yacoubi T, Alraihan J, Alhusain I, Elmokyed F, Alotaibi F, et al. Splenic cystic lymphangioma mimicking hydatid cyst in an adult: a rare diagnostic challenge. Journal of Surgical Case Reports. 2026 Mar;2026(3):rjag174.
5. Murshid MY, AlHawsah AO, AlSayed KT, Ansari FA. Isolated splenic lymphangioma in adulthood. Journal of Surgical Case Reports. 2024 Jun;2024(6):rjae411.
6. Thorat S, Shaji FM. Splenic lymphangioma masquerading as splenic abscess managed by laparoscopic splenectomy: A case report. World Journal of Clinical Oncology. 2023 Oct 24;14(10):440.
7. Pham MD, Nguyen MT, Vo MT, Hoang VT, Pham NT. Splenic lymphangioma in adult patient treated with laparoscopic splenectomy: A rare case report. SAGE Open Medical Case Reports. 2023 Jan;11:2050313X221147196.
8. Kakaje A, Mujahed R, Hamdan O. Isolated splenic lymphangiomas presenting in an infant with isolated anaemia. Case Reports in Medicine. 2020;2020(1):8919424.
9. Efared B, Bako AB, Younssa H, Boubacar I, Zabeirou A, Koura HH, Boureima HS, Amadou S, Coulibaly IS, Lassey JD, Nouhou H. Splenic lymphangiomas as a common indication for splenectomy: a case series with literature review. BMC surgery. 2022 Dec 29;22(1):446.