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Persistent Abdominal Pain Associated with Splenic Lymphangioma in An Adult Patient: A Case Presentation

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Abstract

Splenic lymphangioma is a rare benign lesion, predominantly seen in the pediatric population and exceptionally in adults. It is usually associated with lymphatic system malformations caused by secondary lymphangiectasia due to abnormal communication between lymphatic ducts. It often coexists with complex clinical syndromes, such as Klippel-Trenaunay syndrome, congenital epithelial cysts, or, in less frequent conditions, infectious or post-traumatic triggering events. It typically presents in the neck or axillae, with intra-abdominal cases accounting for less than 5% of all cases. We present the clinical case of a 44-year-old male patient who presented with a clinical course of approximately one month, characterized by progressive abdominal pain associated with nausea, multiple episodes of emesis, anorexia, and involuntary weight loss. A diagnostic laparoscopy was performed, revealing a poorly differentiated lesion in the splenic topography, for which a biopsy was performed, leading to the definitive diagnosis. In this scenario, splenic lymphangioma should be considered among the differential diagnoses of persistent abdominal pain, and definitive interventions should be determined based on clinical characteristics.

Keywords

Lymphangioma, Neoplasms, Splenic Neoplasms, Laparoscopy, Adulthood

1. Introduction

Lymphangiomas are benign malformations of the lymphatic system, usually diagnosed in the pediatric population before the age of 2, and are exceptional in adulthood, typically occurring in individuals under 20 years of age [1] [2]. This pathology is typically located in the neck in 75% to 80% of cases and in the axillary region in 20% of cases, where they are termed cystic hygromas [2] [3]. Intra-abdominal localization is rare, accounting for less than 5% of the cases described in the literature, with the mesentery, retroperitoneum, and omentum being the most common sites [2] [3]. Splenic lymphangioma is particularly atypical, with only 9 cases reported between 1990 and 2010 [2]. Among benign primary splenic tumors, lymphangioma is reported by some authors to occur in just 0.007% of autopsies and surgical interventions [2]-[4].

In the adult population, lymphangiomas are more common in women, with a theorized estrogen-mediated accelerated growth, and their clinical course is generally asymptomatic [2]-[4]. When symptoms do occur, the most common are abdominal pain, anorexia, nausea, emesis, and, in some cases, involuntary weight loss [1]-[6]. However, the diagnosis of lymphangioma in adults presents a challenge due to its predominantly silent clinical course and the low specificity of symptoms when they do present [6]-[8]. This is further complicated by the rarity of the disease in adults, creating a gap in the medical literature and making it difficult to develop evidence-based diagnostic and therapeutic protocols.

Lymphangiomas are histologically classified based on the characteristics of the affected lymphatic ducts, with the main point of differentiation being the size of the ducts. The classification includes three main types: capillary or supermicrocystic, cavernous or microcystic, and cystic or macrocystic [2]-[4]. Their presentation may be solitary, small, subcapsular, or multicystic, which should raise suspicion of other diseases, such as Klippel-Trenaunay syndrome [2]-[5]. Fewer than 300 cases of splenic lymphangioma have been reported in the literature, and reports in Colombia are even more scarce [2] [5].

Regarding imaging characteristics, lymphangiomas are typically identified using ultrasound, computed tomography (CT), or magnetic resonance imaging (MRI) [9]. On ultrasound, lymphangiomas usually appear as anechoic lesions with thin septations and well-defined walls [9] [10]. CT scans may show cystic masses with liquid density, with internal septations that may enhance with contrast [10]. MRI is particularly useful for defining the extent of the lesion and its relationship with adjacent structures, showing lesions with signal intensity similar to fluid on T2 and variable on T1 depending on protein or hemorrhagic content [10] [11]. In the case of splenic lymphangiomas, the imaging presentation is often similar, though they may be confused with other cystic splenic lesions, such as epidermoid cysts, cystic hemangiomas, and cystic neoplasms, highlighting the importance of differential diagnosis [9]-[11].

The differential diagnosis of lymphangioma in adults should include other cystic splenic pathologies, such as splenic cysts, cystic neoplasms, and infectious processes

like splenic abscesses [1]. Additionally, conditions such as Whipple's disease and sarcoidosis, which may manifest with lymphadenopathy and cystic lesions in multiple organs, should be considered [1]-[6]. Accurate differentiation often requires the combination of advanced imaging studies and, in some cases, histopathological confirmation after surgical resection of the lesion [1] [5] [12].

2. Case Presentation

We present the case of a 44-year-old man with no significant medical or surgical history, who presented with a one-month history of left upper quadrant abdominal pain. The pain was lancinating, intermittent, and intensified throughout the day, reaching an intensity of 8/10 on the visual analog scale during exacerbation episodes. In addition to the pain, the patient reported persistent nausea and multiple episodes of food-content emesis, with subsequent relief. No significant changes in bowel habits were noted, but there was a mild sensation of postprandial fullness, possibly related to stomach compression by the splenic mass.

On physical examination, the patient had normal vital signs, a moderate adipose panniculus, present bowel sounds with 11 peristaltic sounds per minute, and tenderness in the left upper quadrant associated with a palpable mass, adherent to deep planes, with irregular borders measuring approximately 1 cm. A contrast-enhanced abdominal and pelvic CT scan was performed, revealing multiple hypodense, possibly cystic images in the splenic topography, some of which were calcified and enhanced after contrast administration (**Figure 1**). A more detailed imaging characterization was obtained through MRI, which did not differ from the CT conclusion (**Figure 2**). Specialized laboratory tests were ordered, showing negative alpha-fetoprotein (2.38 ng/ml; upper limit 10.9 ng/ml), negative carcinoembryonic antigen (0.1 ng/ml; upper limit 4 ng/ml), and negative CA-19.9 (1.6 U/ml; upper limit 40 U/ml).

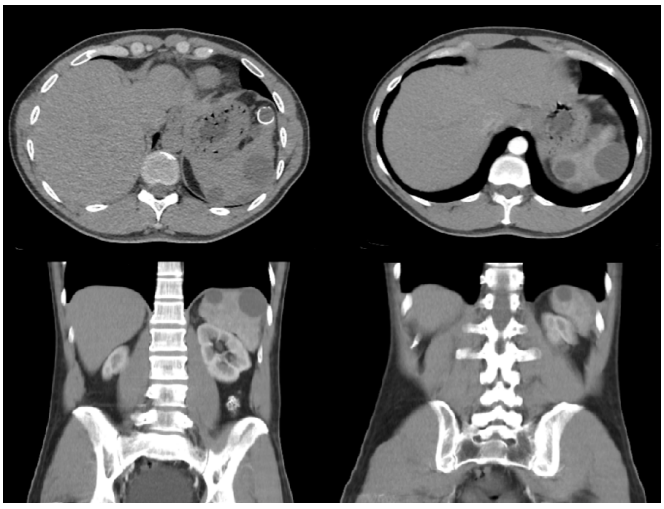

Figure 1. Contrast-enhanced abdominal and pelvic CT: The spleen presents multiple hypodense lesions ranging from 14 to 42 mm, some of which deform the splenic contour, notably one in the anterior aspect with peripheral calcifications.

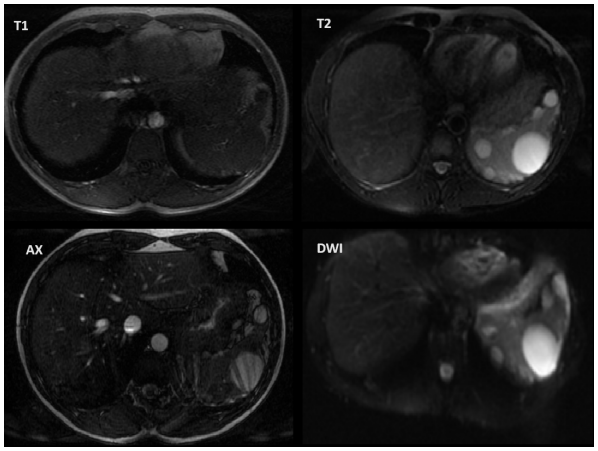

Figure 2. Contrast-enhanced abdominal MRI shows multiple rounded, well-defined, cystic lesions measuring between 10 mm and 40 mm in maximum diameter, with cystic behavior throughout the sequence, and no contrast enhancement observed.

Given the patient's clinical course, imaging, and semiological findings, and considering that the differential diagnosis of splenic lymphangioma includes several entities that may mimic its clinical and imaging presentation, it is essential to consider benign splenic tumors such as hemangiomas, the most common benign splenic neoplasms. Another consideration is the non-parasitic splenic cyst, which presents as a well-defined cystic lesion, generally asymptomatic but capable of causing pain in some cases. Splenic metastases should also be considered, though they are rare and typically associated with primary malignancies in other organs, such as melanoma. Another entity to consider is splenic lymphoma, which typically manifests as diffuse splenomegaly or focal splenic lesions and may present with systemic symptoms such as fever, night sweats, and weight loss. Therefore, a diagnostic laparoscopy was considered for biopsy based on the findings, which were performed without complications. During the intervention, a friable, easily bleeding lesion was found in the lower region of the spleen, with a whitish appearance, ulceration, and signs of fibrosis (**Figure 3**). A biopsy of the lesion was taken, and the patient was sent for post-operative surveillance. Regarding management, considering the benign nature of splenic lymphangioma and the absence of acute complications, a conservative approach to pain management through analgesia was chosen. The importance of regular follow-up to monitor any changes in clinical presentation or lymphangioma characteristics was explained to the patient. The conservative treatment resulted in a favorable outcome. Over the weeks of follow-up, the patient's abdominal pain gradually subsided until it disappeared, as did the emetic episodes. Given this positive evolution and clinical stability, the patient was discharged with recommendations for periodic follow-up.

The pathology report received days later revealed macroscopic and microscopic findings consistent with a benign mesenchymal lesion, characterized by a proliferation of variably sized vascular structures containing lymph, with no evidence of malignancy. Immunohistochemistry studies also showed positivity for D2-40 and CD31, supporting the finding of cystic lesions. The patient continues to show

favorable evolution and is being followed up on an outpatient basis.

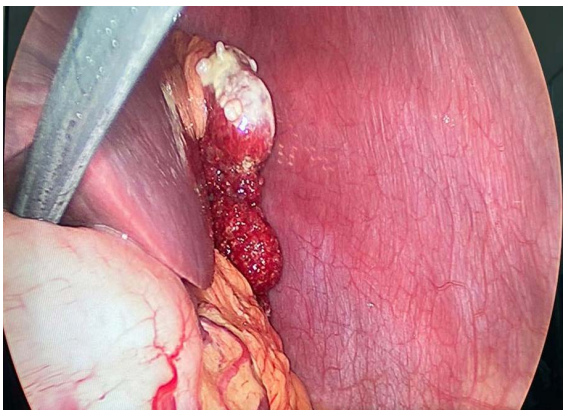

Figure 3. Whitish, ulcerated, stony lesion with signs of fibrosis in the spleen.

3. Discussion

Lymphangiomas are rare lymphatic malformations, typically found in the neck and axillae [1]. These lesions are more commonly observed in the pediatric population, with their occurrence in adulthood being exceptional, representing less than 10% of reported cases [2]. Splenic involvement is even rarer, seen in less than 5% of cases, which underscores the need to consider this entity in the differential diagnosis of patients with chronic abdominal pain, especially when symptoms are nonspecific [2]-[6]. This point is particularly relevant because, although the literature describes a higher prevalence in women, the case presented involves a man, making it an atypical example [1]-[6].

The epidemiology of splenic lymphangiomas is difficult to pinpoint, considering that it is a very atypical presentation, constituting only 0.007% of primary splenic tumors detected in autopsies and surgeries [2]-[4]. These lesions can be isolated or accompanied by other systemic abnormalities, which could suggest the presence of complex clinical syndromes such as Klippel-Trenaunay syndrome [7] [8]. The underlying pathophysiology of lymphangioma formation is believed to be related to lymphatic agenesis leading to lymphangiectasia, a condition that results in the terminal dilation of lymphatic ducts and the subsequent formation of cysts in the affected organ [12] [13].

The clinical presentation of splenic lymphangiomas tends to be nonspecific, complicating their diagnosis [2] [14]. The spectrum of clinical presentations varies from asymptomatic patients, whose diagnosis is incidental, to those with symptoms such as asthenia, adynamia, weight loss, and hyporexia [2]-[5]. Abdominal pain is a common manifestation, often accompanied by atypical symptoms such as referred pain to the shoulder, alterations in bowel habits like constipation or diarrhea, and even fever [15].

The diagnosis of splenic lymphangiomas relies heavily on imaging techniques. Ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI)

are essential for characterizing the lesions [9]. CT and MRI, in addition to revealing cystic formations, can detect peripheral satellite lesions, some of which may be calcified [9]-[11]. Biopsy is crucial for confirming the diagnosis, especially when combined with immunohistochemical markers such as podoplanin (D2-40), a selective marker of lymphatic endothelium, and CD-31, which indicates the endothelial origin of the cells [2] [5]. Other markers such as CD-34, factor VIII, and vascular endothelial growth factor receptor 3 (VEGFR-3) can also be useful in the diagnostic evaluation [2]-[7].

When considering differential diagnoses for splenic cystic lesions, it is essential to distinguish between primary and secondary cysts [11]. Secondary cysts, or pseudocysts, account for about 80% of splenic cystic lesions and generally develop as a result of trauma, infections, or splenic infarction. These cysts are characterized by a fibrous capsule without a specific cellular lining [5] [11]. On the other hand, primary cysts include both parasitic, such as those caused by Echinococcus granulosus (hydatid cysts), which have distinctive histological features like an inner germinal layer, an intermediate laminated membrane, and an external fibrous layer that may contain protoscolices, and non-parasitic ones, which include hemangiomas, epithelial cysts (epidermoid or mesothelial), and lymphangiomas, the latter being the least frequent [2]-[7].

Regarding therapeutic management, complete surgical resection of the spleen is the treatment of choice for splenic lymphangiomas [2] [5]. Although techniques such as cyst drainage or aspiration have been used, their outcomes are suboptimal, which justifies the preference for total splenectomy in cases where it is warranted [2]-[7].

4. Conclusion

Splenic lymphangioma is a rare benign tumor typically observed in the pediatric population, with a much lower incidence in adults. This case demonstrates the importance of considering this condition in the differential diagnosis of abdominal pain, especially when the presentation is nonspecific. Imaging studies and histopathological confirmation are essential for diagnosis. Management should be individualized based on the patient's clinical presentation, and conservative treatment can result in a favorable outcome, as evidenced by this case.

Declaration

Informed consent from the patient and the institution has been obtained for the publication of this case.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Efared, B., Bako, A.B.A., Younssa, H., Boubacar, I., Zabeirou, A., Koura, H.H., *et al.*

(2022) Splenic Lymphangiomas as a Common Indication for Splenectomy: A Case Series with Literature Review. *BMC Surgery*, **22**, Article No. 446. https://doi.org/10.1186/s12893-022-01898-0

[2] Rodríguez-Montes, J.A., Collantes-Bellido, E., Marín-Serrano, E., Prieto-Nieto, I. and Pérez-Robledo, J.P. (2016) Linfangioma esplénico. Un tumor raro. Presentación de 3 casos y revisión de la bibliografía. *Cirugía y Cirujanos*, **84**, 154-159. https://doi.org/10.1016/j.circir.2015.06.016

[3] Perez, A., Perez, M.E.C., Yuga, AC and Viray, B.A.G. (2020) Splenic Lymphangioma in Adulthood: A Case Report. *International Journal of Surgery Case Reports*, **67**, 250-253. https://doi.org/10.1016/j.ijscr.2020.01.061

[4] Al-Shaikh, S.A., Mubarak, A.M. and Harb, Z.F. (2017) Splenic Lymphangioma in an Adult. *Saudi Medical Journal*, **38**, 1148-1152. https://doi.org/10.15537/smj.2017.11.20625

[5] Jiménez-Canizales, C.E., Bolaños, F., Benavides-López, E.F., Mondragón-Cardona, Á.E. and Rojas-Gómez, O.F. (2022) Linfangiomatosis esplénica en un paciente adulto, Reporte de caso. *Acta Medica Peruana*, **39**, 392-395. https://doi.org/10.35663/amp.2022.394.2451

[6] Ioannidis, I. and Kahn, A.G. (2015) Splenic Lymphangioma. *Archives of Pathology & Laboratory Medicine*, **139**, 278-282. https://doi.org/10.5858/arpa.2013-0656-rs

[7] Valderrama, A.R., Pinilla, C.C., Lozano, J.A., Toro, C.A.S., Correa, C.E., Ferez, I.B., and Puyana, A.C. (2002) Linfagioma Esplénico. *Revista Colombiana de Cirugía*, **17**, 174-178. https://www.revistacirugia.org/index.php/cirugia/article/view/1226

[8] Solomou, E.G., Patriarheas, G.V., Mpadra, F.A., Karamouzis, M.V. and Dimopoulos, I. (2003) Asymptomatic Adult Cystic Lymphangioma of the Spleen: Case Report and Review of the Literature. *Magnetic Resonance Imaging*, **21**, 81-84. https://doi.org/10.1016/s0730-725x(02)00624-0

[9] Abbott, R.M., Levy, A.D., Aguilera, N.S., Gorospe, L. and Thompson, W.M. (2004) From the Archives of the AFIP. *RadioGraphics*, **24**, 1137-1163. https://doi.org/10.1148/rg.244045006

[10] Polat, P., Kantarci, M., Alper, F., Suma, S., Koruyucu, M.B. and Okur, A. (2003) Hydatid Disease from Head to Toe. *RadioGraphics*, **23**, 475-494. https://doi.org/10.1148/rg.232025704

[11] Thipphavong, S., Duigenan, S., Schindera, S.T., Gee, M.S. and Philips, S. (2014) Nonneoplastic, Benign, and Malignant Splenic Diseases: Cross-Sectional Imaging Findings and Rare Disease Entities. *American Journal of Roentgenology*, **203**, 315-322. https://doi.org/10.2214/ajr.13.11777

[12] Beltrán, M.A., Barría, C., Pujado, B., Oliva, J., Contreras, M.A., Wilson, C.S., *et al.* (2009) Linfangioma esplénico gigante: Caso clínico. *Revista Médica de Chile*, **137**, 1597-1601. https://doi.org/10.4067/s0034-98872009001200008

[13] Chung, S.H., Park, Y.S., Jo, Y.J., Kim, S.H., Jun, D.W., Son, B.K., *et al.* (2009) Asymptomatic Lymphangioma Involving the Spleen Andretroperitoneum in Adults. *World Journal of Gastroenterology*, **15**, 5620-5623. https://doi.org/10.3748/wjg.15.5620

[14] Yang, F. (2013) Splenic Lymphangioma That Manifested as a Solid-Cystic Mass: A Case Report. *World Journal of Gastroenterology*, **19**, 781-783. https://doi.org/10.3748/wjg.v19.i5.781

[15] Vezzoli, M., Ottini, E., Montagna, M., La Fianza, A., Paulli, M., Rosso, R. and Mazzone, A. (2000) Lymphangioma of the Spleen in an Elderly Patient. *Haematologica*, **85**, 314-317.