International Surgery Journal
Marques S et al. Int Surg J. 2024 Dec;11(12):2126-2130
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20243559

Case Report

A rare and fortunate case of unruptured splenic peliosis – case report and literature review

Sónia Marques[1]*, Natacha Nunes[1], Lara Madeira[1], Renato Barradas[1], Paulo Sintra[1], Miguel Rodrigues[2]

[1]Department of General Surgery, Unidade Local de Saúde da Lezíria, Santarém, Portugal
[2]Department of Pathology, Unidade Local de Saúde da Lezíria, Santarém, Portugal

Received: 01 November 2024
Accepted: 18 November 2024

***Correspondence:**
Dr. Sónia Marques,
E-mail: soniamarques224@hotmail.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

Splenic peliosis is an extremely rare pathological entity of unclear etiology characterized by the presence of multiple blood-filled cavities. Although numerous associations have been described in the literature, the etiology remains unclear. The present case is about a healthy 40-year-old woman who was referred to our institution due to chronic symptoms of abdominal discomfort and constipation. Despite the abdominal computed tomography (CT) scan and abdominal magnetic resonance imaging (MRI) reported an adrenal mass, reviewing the scans, the radiologist suggested a 17,5 cm, well-vascularized splenic mass. A splenectomy was performed, without any complications. We discuss the current available literature and emphasize the importance of awareness of this diagnosis.

Keywords: Isolated peliosis, Spleen, Splenectomy, Unruptured peliosis, Uncommon splenic diseases, Splenic tumour-like lesions

INTRODUCTION

Peliosis is an extremely rare pathological entity with fewer than 100 documented cases in medical literature. Peliosis presents as a non-neoplastic condition marked by the development of blood-filled cystic spaces within parenchymatous organs.[1,2]

The underlying pathophysiological mechanism appears to involve localized microcirculatory disruptions, specifically affecting sinusoidal wall integrity and regional blood pressure dynamics.[3] The condition's nomenclature stems from the Greek term "pelios," first employed by Wagner in 1861 to document hepatic abnormalities. Subsequently, in 1866, Cohnheim documented the first autopsy-confirmed case of splenic involvement.[1]

While splenic peliosis typically coincides with hepatic manifestations, isolated splenic involvement represents an even more exceptional occurrence. These lesions typically remain clinically silent, often discovered as incidental findings during computed tomography examinations or post-mortem studies.[1,4] Nevertheless, cases can manifest dramatically through spontaneous splenic rupture, precipitating life-endangering hemorrhagic shock that necessitates immediate surgical management.[4,5]

Despite extensive documentation of various associations with pharmaceutical agents and medical conditions in the scientific literature, the precise etiology remains incompletely understood.[4] Current evidence suggests links with several chronic conditions, encompassing malignancies, human immune-deficiency virus (HIV) infection, tuberculosis, and hepatic cirrhosis, as well as associations with pharmacological agents including anabolic steroids and oral contraceptives.[5,6]

We report a case of an adult female patient with a significant splenomegaly caused by isolated splenic peliosis. Pathological, radiological and surgical findings

along with the management plan for the patient are discussed.

CASE REPORT

A healthy 40-year-old woman was referred to our institution with complaints of abdominal discomfort, postprandial bloating and fullness, and constipation for months, without any complains of weight loss, asthenia, anorexia. The patient had no personal history and the family history was negative for inherent disease. At physical examination, she experimented discomfort in the upper right quadrant, with no abdominal tenderness, and no evident mass. Although the abdominal computed tomography (CT) scan and abdominal magnetic resonance imaging (MRI) reported an adrenal mass, after reviewing the scans, the radiologist suggested a giant, well-vascularized splenic mass with no other lesions in the abdomen (Figures 1a, 1b, and 2a).

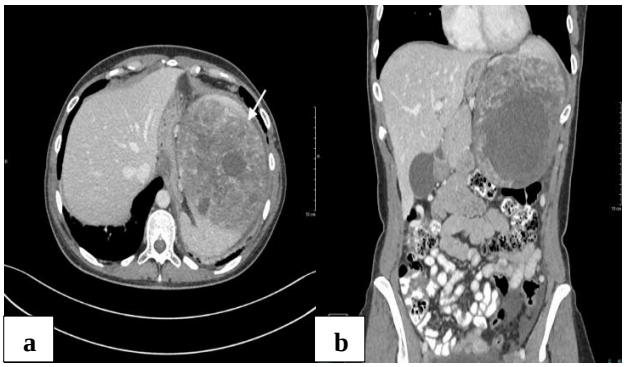

Figure 1: Contrast-enhanced computed tomography scan (a) axial view and (b) coronal view. Significant splenomegaly (17.5×15×9 cm) with solid components and hypodense cystic areas consistent with peliosis.

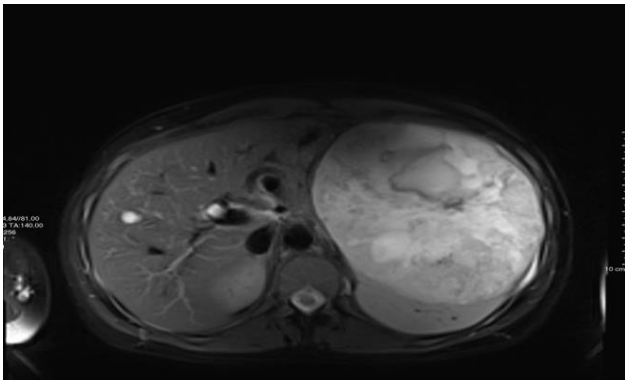

Figure 2: T2-weighted magnetic resonance images. Enlarged spleen with a heterogeneous appearance, with solid areas, cystic formations and a discrete hemorrhagic component.

The surgical team discussed these findings with the patient and a shared decision-making to a laparotomic diagnostic approach was made with the aim to relieve the symptoms and obtain a final diagnosis. No abnormalities were found on the laboratory examinations, including complete blood cell count, coagulation profile and tumoral markers. 2 weeks previous the surgery the patient was vaccinated with pneumococcal, meningococcal and haemophilis influenza type b vaccines to optimize the immune response and, consequently, decrease the risk of overwhelming post-splenectomy sepsis. She was admitted to our surgical unit the previous day and received a perioperative medical checkup. On the second day of hospitalization, the scheduled operation was performed. The surgical approach was a left subcostal laparotomy due to the significant splenomegaly. Exploration of the entire abdominal cavity showed an enlarged spleen. There was no evidence of active or ongoing bleeding on the spleen or clotted blood in the peritoneal cavity. A splenectomy was performed, without any complications (Figure 3). Operative time was 100 minutes. The patient had an uneventful recovery and was discharged home on the 4th postoperative day.

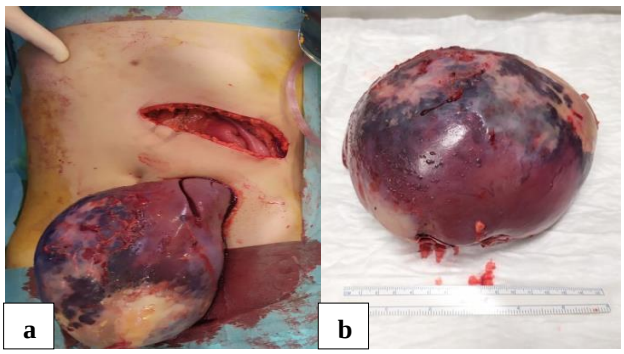

Figure 3 (a and b): Explorative left subcostal laparotomy showing a significant splenomegaly with heterogeneous appearance and nodular surface, with reddish-purple lesions shining through the capsule.

Macroscopic examination showed a spleen weighing 1098 g and measuring 17.5×15×9 cm, diffusely haemorrhagic with a peripheral rim of preserved parenchyma. Cystic areas filled with haemorrhagic content, varying in size between 1 and 8 cm, were identified. Additionally, areas of fibrin and necrosis were noted (Figure 4).

Figure 4: Macroscopic appearance of splenic peliosis.

Histological examination revealed that the haemorrhagic lesion consisted of multiple interconnected cavities and cyst-like structures, mostly devoid of their epithelial lining and filled with erythrocytes and fibrin. Areas of coagulative necrosis were present. At the periphery of the lesion, there was a thin rim of splenic parenchyma with preserved general architecture, atrophy of the white pulp, and slight dilation of the sinuses (Figure 5a-c).

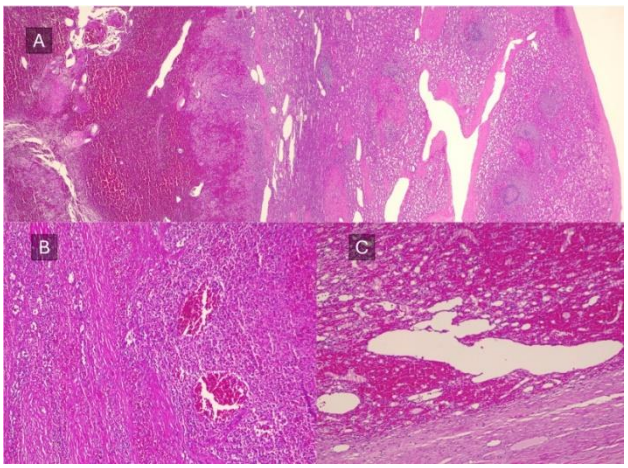

Figure 5: (a) (H&E, 2.5x) – Low power view of the spleen, showing a transition between the somewhat preserved spleen, with atrophic white pulp and slight dilation of the sinuses, and the presence of multiple dilated cysts and haemorrhagic parenchyma with associated fibrin; (b and c) (H&E, 10x) – high power view of the cysts with no epithelial lining and with endothelial lining.

No microorganisms were identified. A diagnosis of splenic peliosis was rendered.

The patient was discharged from the surgery appointment and keeps being followed by the family doctor. To date, three years postoperative ultrasounds have shown no evidence of other organs being involved and the patient remains well.

DISCUSSION

The vascular condition known as peliosis manifests through the abnormal dilation of blood vessels within solid organs, culminating in the formation of blood-containing cystic spaces. This pathological process predominantly affects organs associated with the mononuclear phagocytic system, including hepatic tissue, splenic parenchyma, bone marrow, and abdominal lymph nodes.[4,7,8] Splenic involvements rarely occurs in isolation, more commonly presenting as a secondary manifestation to hepatic peliosis. Isolated splenic peliosis represents an exceptionally uncommon variant of this condition.[5] In our institutional experience, we have documented a single case of isolated splenic peliosis, which represents the second such case reported in Portugal.[9] A comprehensive review of English-language medical literature reveals only 48 (n=48 cases) documented cases of isolated splenic peliosis. Analysis of these cases demonstrates that spontaneous rupture constituted the primary presentation in 66% of patients (n=32/48), while traumatic events accounted for 6% (n=3/48) of cases. The remaining 28% (n=13/48) were discovered either incidentally or during evaluation of mild abdominal discomfort.[1,4,5]

The pathophysiological basis of this condition remains largely unknown. Multiple hypotheses have been proposed regarding its underlying mechanisms. One theory suggests that the process initiates with parenchymal necrosis, followed by sinusoidal distension in response to elevated intravascular pressure. Alternative postulations characterize the condition as an acquired vascular anomaly, attributing its development either to sinusoidal wall damage or to active vessel proliferation resembling benign vascular neoplasms. Further contributing to the complexity of potential mechanisms, several investigations have noted the presence of immune complex deposits in proximity to the affected areas.[1,9]

While the precise causative factors remain incompletely understood, a wide variety of underlying conditions has been documented in association with peliosis development. These encompass various chronic debilitating disorders, including tuberculosis, malignant conditions, prolonged alcohol abuse, hepatic cirrhosis, viral hepatitis (types B and C), renal insufficiency, diabetes mellitus, AIDS, post-transplantation immunosuppression, and chemotherapy-induced states. Pharmaceutical agents have been particularly implicated, with the most frequent associations observed in patients receiving long-term treatment with anabolic androgenic steroids, danazol, or azathioprine, among other medications.[1,7,8]

Hematologic disorders represent another significant category of associated conditions, with documented cases involving multiple myeloma, aplastic anemia, Hodgkin disease, and myelofibrosis.[4,10] Recent research has suggested a potential mechanistic link to dysregulated angiogenesis, specifically through elevated vascular endothelial growth factor (VEGF) levels, a crucial mediator of normal angiogenic processes.[4] In some cases, no predisposing factor could be determined.[1] In our patient the only predisposing factor that can be associate is the intake of oral contraceptive pills.

The clinical presentation of splenic peliosis exhibits considerable variability, with many individuals remaining asymptomatic until the condition is discovered incidentally through imaging studies, during post-mortem examination, or following spontaneous organ rupture, which may occur with minimal or no traumatic trigger.[4,11] Clinical signs are variable and non-specific, ranging from complete absence of symptoms to non-specific abdominal discomfort, potentially progressing to life-threatening hemorrhagic complications in cases of splenic rupture. Many affected individuals only seek medical evaluation

after developing significant organ enlargement or experiencing rupture-related symptoms. Documented cases span a broad age spectrum, encompassing patients from adolescence through advanced age (14-82 years), without demonstrating any significant predilection for either gender.[1,10] Our patient was a 40-year-old woman that was referred to our institution due to chronic symptoms of abdominal discomfort, post-meal bloating and fullness, accompanied by altered bowel habits manifesting as constipation.

Multiple imaging techniques may be employed for splenic evaluation, with the selection typically guided by clinical presentation and diagnostic considerations. These diagnostic modalities encompass ultrasonography, computed tomography, PET-CT, and magnetic resonance imaging.[4] Characteristic radiological features typically reveal multiple diminutive hypodense lesions with cystic characteristics. In cases of cystic rupture, imaging may demonstrate subcapsular hematoma formation, parenchymal laceration, or intraperitoneal hemorrhage. On ultrasound examination, these lesions typically display hyperechogenicity, with their structural appearance varying between heterogeneous and homogeneous patterns, depending on the presence of necrotic changes or hemorrhagic components. Larger lesions characteristically exhibit moderate posterior acoustic enhancement. Computed tomography typically reveals well-circumscribed, multiloculated hypodense areas, which may demonstrate increased density in the context of hemorrhage. The lesions generally lack calcification and remain confined within the capsule unless rupture occurs, whereupon subcapsular hematoma formation may accompany intraperitoneal blood collection. Magnetic resonance imaging findings demonstrate variability, though T2-weighted sequences typically reveal hyperintense signals attributable to the cystic contents.[1,11,12] Despite these various modalities, imaging may be inadequate for establishing a clear diagnosis as in the case of our patient.[4]

Gross anatomical examination of affected tissue typically demonstrates numerous cavitous structures filled with blood throughout the splenic parenchyma. These spaces exhibit round to ovoid morphology and can achieve diameters of several centimeters, becoming visible both through the capsular surface and within the parenchymal tissue. The distributional pattern of these cavities shows considerable variation, appearing either in sporadic isolation, irregular clusters, or diffuse organ involvement. In our specimen, cavities ranged from 1 to 8 cm in diameter and were distributed throughout the whole organ. Histologically, two different types of lesions have been described. The first, termed "phlebectatic peliosis," is characterized by cavities featuring endothelial lining and/or fibrotic walls. The second variant, designated as "parenchymal peliosis," exhibits cavities devoid of cellular lining.[1] Our pathological analysis revealed the presence of both architectural patterns, with a predominance of the parenchymal variant.

The radiological differential diagnosis encompasses several entities requiring careful consideration. These include lymphoproliferative disorders (particularly small cell and splenic marginal zone lymphomas), vascular neoplasms (such as hemangiomas, angiosarcomas, and splenic hamartomas), infectious processes (including abscess formation and tuberculous involvement), and post-traumatic cystic formations. From a histopathological perspective, the diagnostic considerations must distinguish this entity from other conditions that can produce similar morphological patterns. These include secondary congestive changes associated with veno-occlusive disease, vascular neoplasms such as hemangiomas, and infiltrative processes like hairy cell leukaemia.[1,9,12]

The optimal surgical approach for incidentally discovered splenic peliosis remains undefined, with no established treatment guidelines. Given the spleen's crucial immunological and endocrine functions, the decision to proceed with total splenectomy requires careful consideration of risk-benefit ratios. While some clinicians advocate prophylactic splenectomy for incidentally diagnosed cases, particularly in patients in high risk of rupture, as this approach provides definitive histological diagnosis and eliminates future hemorrhage risk, this recommendation remains controversial.[13] Management strategies differ based on clinical presentation and hemodynamic status. In cases of spontaneous rupture with hemodynamic instability, immediate intervention is mandatory, necessitating massive transfusion protocol activation and emergency splenectomy via laparotomy. If hemodynamically stable, they often undergo semi-elective open or laparoscopic splenectomy. Although splenic arterial embolization is frequently suggested as a potential therapeutic option, documented evidence of its application in this context remains sparse. The post-operative course may be complicated by pleural effusions and intra-abdominal fluid collections.[4]

Standard algorithms for the follow-up of patients with isolated splenic peliosis do not exist. The documented association between this condition and hematologic malignancies warrants particular attention, necessitating clinician vigilance regarding potential concurrent malignant processes.[4] Healthcare providers should also exercise caution when considering the prescription of medications previously linked to peliosis development.[14] In our reported case, the management strategy included counseling the patient to discontinue oral contraceptive use and explore alternative contraceptive methods. Additionally, she was also referred to a hematology appointment. The three-year post-operative assessment revealed no evidence of peliotic involvement in other organs, and the patient maintains good health at present.

CONCLUSION

Isolated unrupted splenic peliosis with no comorbidities is an extraordinary rare condition and frequently overlooked as a potential differential diagnosis in cases of idiopathic

splenomegaly. The limited number of documented cases presents a significant obstacle in developing standardized treatment protocols. When evaluating splenic masses, clinicians must maintain awareness of peliosis as a diagnostic possibility, given its potential for catastrophic outcomes if unrecognized. Total splenectomy emerges as the most judicious therapeutic approach in these cases, considering that splenic rupture, should it occur, can precipitate life-threatening intraperitoneal hemorrhage.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Blichárová A, Benetinová Z, Mátyás T, Labaj P, Verbóová Ľ, Tancoš V, et al. Peliosis of the spleen as an unusual cause of splenic rupture: A case report and a review of literature. J Forensic Leg Med. 2024;103:102659.
2. Adachi K, Ui M, Nojima H, Takada Y, Enatsu K. Isolated splenic peliosis presenting with giant splenomegaly and severe coagulopathy. Am J Surg. 2011;202(2):e17-9.
3. Junior M, Zanco G, Sala M, El-Ibrahim R. Splenic peliosis: a case report. Braz Arch Digest Surg. 2011;24(2).
4. Chandra A, Navarro SM, Killeen TF, Nemanich G, Harmon JV. Spontaneous splenic rupture due to Peliosis and the association with malignancy: A case series and literature review. Int J Surg Case Rep. 2023;110:108676.
5. Nassr B, Hassan W, Nassr HN, Allouzi A, Al-Shebly M. Isolated splenic peliosis: a case report. J Med Case Rep. 2023;17:298.
6. Terada T. Isolated splenic peliosis: Case report and consideration of histogenesis. Human Pathol Case Rep. 2015;144:1.
7. Mohandas P, Krim AOA, Samson P. Unexpected finding of splenic peliosis in a traumatic spleen in a patient with cleidocranial dysplasia. SA J Radiol. 2018;22(1):1371.
8. Magwai MG, Andronikou S. Isolated splenic peliosis in an immunocompromised patient. S Afr J Surg. 2012;50(3):92.
9. Santos B, Mendez B, Almeida P, Dias B, Miguel I, Pina S, et al. Spontaneous splenic rupture in the context of taking DOAC in a patient with splenic peliosis. Revista Algarve Médico. 2022;20.
10. Begum S, Muhammad K. Splenic Peliosis and Rupture − A Surgical Emergency Case Report and Review of the Available Literature. J Appl Hematol. 2016;7(4):143-7.
11. Abo D, Terae S, Fukasawa Y, Seki H, Hasegawa Y, Sakuhara Y, et al. Splenic peliosis mimicking a solid tumor: findings in multimodalities. J Comput Assist Tomogr. 2008;32(6):890-2.
12. Davidson J, Tung K. Splenic peliosis: an unusual entity. Br J Radiol. 2010;83(990):e126-80.
13. Rhu J, Cho J. Ruptured splenic peliosis in a patient with no comorbidity: A case report. World J Clin Cases. 2020;8(3):535-9.
14. Lashbrook DJ, James RW, Phillips AJ, Holbrook AG, Agombar AC. Splenic peliosis with spontaneous splenic rupture: report of two cases. BMC Surg. 2006;6:9.

Cite this article as: Marques S, Nunes N, Madeira L, Barradas R, Sintra P, Rodrigues M. A rare and fortunate case of unruptured splenic peliosis – case report and literature review. Int Surg J 2024;11:2126-30.