

## Case Report

# Spontaneous Rupture of the Spleen on a Patient with Splenic Hemangioma and Multiple Cystic Lesions in Kidney and Liver. Case Report and Literature Review

Andreea Iliesiu<sup>1,2</sup>, Oana Cristina Ciornenchi<sup>1</sup>, M. Tampa<sup>2,3</sup>, Clara Matei<sup>2</sup>,  
Simona-Roxana Georgescu<sup>2,3</sup>, C. Cirstoiu<sup>1,2</sup>, N. Zarnescu<sup>1,2</sup>, Maria Sajin<sup>1,2</sup>, A. Dumitru<sup>1</sup>

<sup>1</sup>University Hospital of Emergencies, Bucharest, Romania

<sup>2</sup>“Carol Davila” University of Medicine and Pharmacy Bucharest, Romania

<sup>3</sup>Department of Dermatology, “Victor Babes” Hospital for Infectious Diseases

### REZUMAT

*Ruptura spontană a splinei la un pacient cu hemangiom splenic asociat cu multiple leziuni chistice la nivel hepatic și renal. Prezentare de caz și review al literaturii medicale în domeniu*

**Obiective:** Ruptura atraumatică de splină este o entitate clinică rar întâlnită, cu etiologie plurifactorială. Scopul acestui articol este de a prezenta cazul rar al unui pacient asimptomatic cu hemangiom splenic de mărime medie, ce asociază multiple alte leziuni chistice la nivel hepatic și renal și, de asemenea, să treacă în revistă literatura medicală în domeniu.

**Prezentarea cazului:** Prezentăm cazul unui pacient de sex masculin în vârstă de 33 de ani, fără istoric medical relevant, care se prezintă la unitatea de primiri urgențe cu durere abdominală și paloare intensă. În urma laparotomiei exploratorii de urgență cu splenectomie, au fost decelate hemoperitoneu masiv și lacerări ale splinei. Piesa recoltată a fost trimisă spre examinare anatomopatologică.

**Rezultate:** Examinarea histologică a evidențiat lacune vasculare de mărimi variate, delimitate de un singur strat de celule endoteliale cu nucleu ce proemină în lumen, fără mitoze, separate de trabecule fibroase. Diagnosticul de hemangiom splenic a putut fi astfel confirmat; investigațiile imagistice au evidențiat, de asemenea, leziuni chistice multiple la nivel hepatic și renal.

**Concluzii:** Acest caz este particular întrucât pacientul a dezvoltat complicații severe ale hemangiomului splenic fără simptome prealabile, și pentru că leziunea splenică asociază prezența de leziuni chistice. Din acest motiv, articolul include o trecere în revistă a literaturii medicale în domeniul sindromelor care asociază prezența de hemangioame cu localizare la nivelul splinei.

**Cuvinte cheie:** ruptură splenică, hemangiom, chist

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**Corresponding author:**

Prof. Simona-Roxana Georgescu, MD, PhD

Department of Dermatology, “Carol Davila” University of Medicine and Pharmacy

37 Dionisie Lupu Street, Bucharest, Romania

e-mail: simonaroxanageorgescu@yahoo.com

## ABSTRACT

**Objectives:** A traumatic splenic rupture is a rare clinical entity with multiple possible underlying etiologies. The aim of this article is to report a case of unusual presentation of an average size splenic hemangioma of an asymptomatic patient with multiple other cystic lesions in kidneys and liver and to summarize the syndromes that include this pathologic entity.

**Report of case:** We present a case of a 33 years old Caucasian male patient with no medical history known who presented at E.R. with acute abdominal pain and intense pallor. Emergency laparotomy was performed. Intraoperatory a massive hemoperitoneum and laceration of the spleen was found; splenectomy was performed. The surgical specimen was histologically analyzed.

**Results:** The histological examination revealed vascular spaces of irregular size delimited by a single layer of blood endothelial cells with nuclei bulging into the lumen, without mitoses, separated by fibrous trabeculae. Diagnosis of splenic hemangioma was confirmed. Imagistic also found multiple cystic lesions in kidneys and liver.

**Conclusions:** The case is notable because the patient was asymptomatic and developed the most severe complication of the splenic hemangioma and also because cystic lesions in other sites were associated. This is the reason why we made a literature review of syndromes which includes splenic hemangioma.

**Keywords:** splenic rupture, hemangioma, cystic lesion

## INTRODUCTION

Splenic hemangioma is a vascular malformation which is one of the most common benign neoplasms of the spleen. They may represent small, incidental lesions that can produce significant splenomegaly and predispose to splenic rupture. These tumors are rare and resemble their counterparts in other organs. The vascular spaces are usually irregular and cavernous, containing abundant red blood cells. Differential diagnoses include benign or malignant neoplasms, metastases, lymphoma, splenic abscess or cystic lesions. The incidence of autopsy examination of their diagnosis ranges from 0.03 to 14% [1], and most often is encountered in adults between the ages of 30 and 50 [2]. Most are small lesions, usually clinically silent, diagnosed accidentally. Sometimes, however, they may be symptomatic, manifested by splenomegaly, abdominal pain, bowel disorders, anemia and thrombocytopenia, Kasabach-Merritt syndrome (anemia, thrombocytopenia and coagulopathy) and in rare cases by spontaneous rupture of the spleen [3].

Splenic rupture is almost always traumatic. In rare cases, when no injury can be incriminated, the etiology may be due to a viral infection (especially mononucleosis), amyloidosis, hematologic malignancies, and rheumatoid arthritis [4]. Spleen rupture can also be associated with a hemangioma, which is

the most common benign tumor of the spleen [3].

The main aim of this article is to present the potential impact on vital prognosis of a benign pathological entity and to mark the association of splenic hemangioma with other multiple cystic lesions in the liver and kidney (imagistic diagnosed).

## CASE REPORT

We present the case of a male patient, aged 33 years, brought by the ambulance to the emergency room after being found fallen in the street. Symptoms at presentation were typical of acute surgical abdomen with signs of peritoneal irritation, diffuse abdominal pain and intense pallor; the patient has not presented fever, jaundice, nausea, vomiting, or had a history of melena or hematuria. Emergency routine exams were performed; the complete blood count (CBC) showed no anemia or thrombocytopenia (Hb - 13g/dl, platelets 220,000/mm<sup>3</sup>), with 20,000 leukocytes and neutrophils (86%), biochemistry was in normal parameters of transaminases; urinalysis without hematuria, leucocyturia or nitrozoeria.

Abdominal ultrasound revealed an increased spleen size and at the inferior pole multiple hypoechoic images located intraparenchymal that presented subcapsular halo. Abdominal fluid in low/medium volume and floating bowel loops were also noted. For a more accurate diagnosis a CT thoraco-

abdomino-pelvic was performed which confirmed the diagnosis of splenomegaly and splenic hemangioma with spontaneous rupture in the absence of any traumatic changes (increased spleen size 15.6/4.4 cm in the axial plane and 13 cm cranial-caudal contours irregular and heterogeneous structure especially in the lower pole by the presence of a large hypodense area of approximately 7 cm; without posttraumatic changes in the parenchymal organs and abdominal-pelvic cavity without pleural or pericardial effusions without posttraumatic bone structural changes) and cystic lesions of liver and kidney (two essential cysts located in the liver in segments IV, V, multiple bilateral renal cysts, most with dimension around 1 cm, max 2.7 cm located on the right lower pole).

An emergency laparotomy with splenectomy in hilum was chosen; hemoperitoneum was drained (about 1.5 l blood) and spleen was histologically examined.

#### Positive diagnosis

On gross examination we identified the spleen with dimensions of 16/11/3 cm, continuity solution that extends from the splenic hilum (**Fig. 1**) to the diaphragm face for a 14 cm length; the capsule was smooth with an uniform appearance of splenic parenchyma and elastic consistency. On the section at the level of the splenic hilum multiple foci of hemorrhage and the presence of diffuse intra-parenchymal subcapsular organized hematoma were found.

Under the microscope we observed small vascular channels, delimited by endothelium with round-oval nuclei that bulge into the vascular lumen

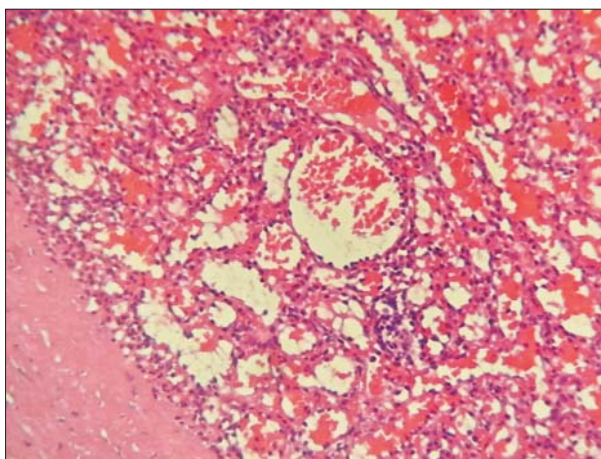


Figure 2.

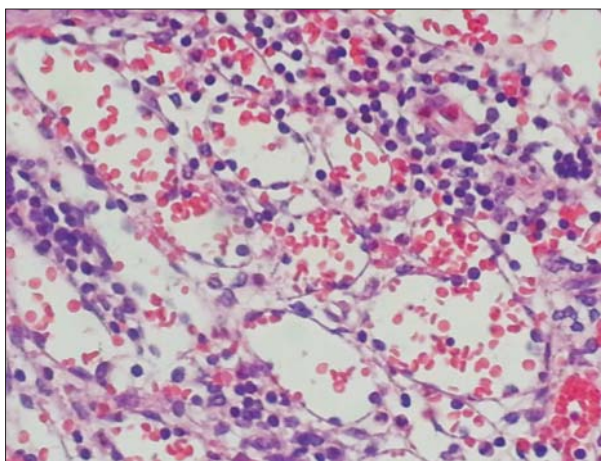


Figure 3.

(**Fig. 4**) and diffuse areas of fibroblast proliferation (**Fig. 3**). It was also noticed that the splenic red pulp is partially occupied by benign proliferation and



Figure 1.

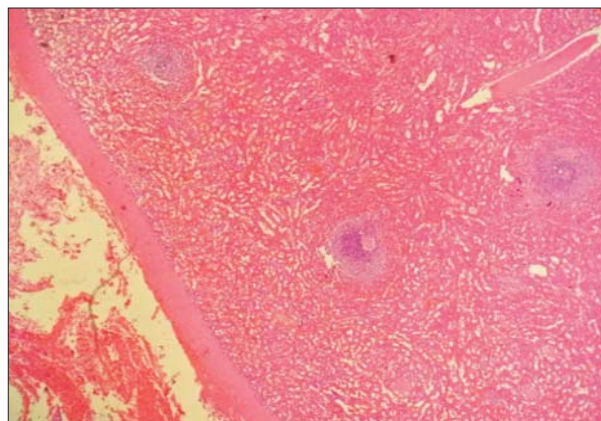


Figure 4.



focal white pulp was atrophic in appearance (Fig. 2). We did not identify areas of necrosis or nuclear or mitotic atypia, and we established the diagnosis of splenic capillary hemangioma.

Endothelial cells in most of these lesions vary in shape from flattened to cuboidal and express CD34, von Willebrand factor and Ulex europaeus lectin but not CD 8. This immunophenotype is indicative of origin from non-sinusoidal endothelium. The most extensive examples, involving almost all of the splenic parenchyma, have been categorized as cases of diffuse angiomatosis. Origin from sinusoidal endothelium (CD 8 positive) has not been specifically excluded in most reported cases of diffuse angiomatosis and there may be an overlap with littoral cell angiomias.

### Treatment

In most cases splenic hemangiomas are benign lesions with a slow growth rate. Hemangiomas are generally not treated unless they are symptomatic (digestive symptoms or modifications of CBC) or very large, with increased risk of hemorrhage; treatment then usually consists in a splenectomy. In this case, although asymptomatic and medium in size, the hemangioma could severely affect life prognosis, and, the splenectomy was performed without any previous symptoms before the spontaneous rupture of the organ.

### DISCUSSIONS

The spleen is an organ composed of red heavily vascularized pulp (which forms the majority of splenic parenchyma and is headquarters phagocytosis) and white pulp, which is the lymphoid component; physiologically, the spleen has many functions, including filtering, capturing aged erythrocytes, extramedullary hematopoiesis and immune response to bacterial aggression [5]. The spleen is susceptible to a wide range of diseases: infectious and inflammatory, vascular and hematologic malignancies. Classically, they are divided according to the affected parenchyma. White pulp causes various diseases including reactive hyperplasia (rheumatoid arthritis, autoimmune thrombocytopenic purpura, AIDS, etc.), malignant lymphoma, and lymphoproliferative diseases (CLL, B-cell lymphoma, T-cell lymphoma and Hodgkin's lymphomas). Disorders that have predilection for the red pulp include infectious diseases (mononucleosis), congestive

heart failure (hemolytic anemia), teaurismoses (Gaucher disease), leukemia (chronic myelogenous leukemia, hairy cell leukemia) non hematopoietic tumors, hamartomas, lymphangioma, angiosarcomas and metastasis) [6].

Splenic hemangioma was first described by Virchow in 1863; no more than 100 cases were described in the medical literature until the year 2000 [1]. Although the etiology of spleen hemangiomas is still unknown, it has been suggested that it is a congenital nevus that may or may not grow in size to become symptomatic [7,8]. Due to the slow growth of this tumor, most cases are diagnosed after the third decade of life, but cases have been described in the pediatric population in the context of various congenital syndromes. Clinical manifestations (abdominal pain, anemia, ascites, weight loss, thrombocytopenia, and the most dreaded of all, spontaneous spleen rupture) of this condition appear to be directly proportional to tumor size.

Although the case presented above illustrates a noisy manifestation with potentially fatal evolution of a rare pathological entity, the lack of any hematologic dyscrasia or other clinical manifestations is what gives an additional particularity of the case. Blood count on admission to hospital did not show anemia, leucopenia or thrombocytopenia. Hepatitis or systemic etiology of congestion as the cause of splenomegaly were excluded by completely normal biochemistry and imaging. Cultures of biological fluids (peritoneal fluid, blood, urine culture) and the lack of negative lymph nodes excluded an infectious cause of splenomegaly. Hematologic and imaging exams were also eliminated as a possible etiology of hematological malignancy/nodes. The decision for exploratory laparotomy was taken by the emergency of stopping the bleeding. Splenic rupture was diagnosed intraoperative. Histological examination diagnosed splenic capillary hemangioma.

It is also interesting to note that the patient was also diagnosed with kidney and liver cystic formation, by imaging techniques; therefore we considered appropriate reviewing syndromes of employing these pathological entities:

Klippel-Trenaunay syndrome: congenital abnormality of the blood vessels characterized by the clinical triad: developing varicose superficial vessels, vascular stains and port wine-colored bone or soft tissue hypertrophy localized unilaterally, usually involving the extremities. The complete triad is found in 63% of cases, but for diagnostic are sufficient two

criteria. This syndrome is associated with visceral vascular malformations including hemangiomas and lymphangioma in colon, small intestine, bladder, kidney, spleen, liver and CNS organs. The most common complications are stasis dermatitis, coagulopathy, pulmonary embolism, heart failure, bleeding from blood vessels; the clinic manifestation are melena, hematemesis and hematuria. Mortality of these patients is situated around 1% [9-12].

Rendu-Osler-Weber syndrome: autosomal dominant hemorrhagic telangiectasia characterized by abnormal vascular architecture in favorite sites [13]. Bleeding is due to vascular abnormalities and bleeding syndromes; their etiology is incompletely elucidated. Clinic manifestation includes bleeding manifested by epistaxis, gingival bleeding, hemoptysis or bleedings with gastrointestinal origin. The skin and frequently present pulsatile vascular papules of various sizes, which can coexist with hemangiomas localized in CNS, lungs, gastrointestinal tract, hepatic and splenic [13].

#### *Proteus syndrome*

The etiology is not fully elucidated, but it is hypothesized as a somatic mutation of a dominant gene, which is typically fatal, unless the mosaicism is present [14]. Patients with this syndrome are apparently healthy at birth, developing specific clinical signs during their growth; the diagnosis is usually made in adolescence [14].

Wideman-Beckwith syndrome is associated with high risk of developing parenchymal neoplasia [15]. Clinical appearance is characterized by organomegaly. Macroglossia (most common symptom), omphalocele, hemihypertrophy of the limbs and paired organs, pancreatic islet hyperplasia, nephromegaly, hepatosplenomegaly make the full clinical picture of this syndrome [15].

In literature cases of both liver and spleen hemangioma coexistence in female patients in the context of prolonged administration of estrogen-progestogen medication have been reported [16]. Splenic hemangiomatosis postpartum [17] as well as giant splenic hemangioma were also reported in the absence of underlying liver diseases [18].

## CONCLUSIONS

This case is not only a review of a rare pathology but also highlights the potential impact on vital prognosis of a benign pathologies. Presented patient

had no medical history, was completely asymptomatic and yet has developed the most severe complication of splenic hemangioma. Therefore, when abdominal ultrasound is done regardless of circumstance should pay increased attention on the spleen. Even without blood count changes or a history of infections, hematologic malignancies, systemic congestion conditions, portal hypertension, etc. the doctor who sees a patient with splenomegaly should take account of such pathology, remaining to be determined appropriateness of treatment (splenectomy).

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## Conflict of interests

None to declare.

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