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LETTER TO THE EDITOR / ПИСМО УРЕДНИКУ

Spontaneous splenic rupture in marginal zone lymphoma at presentation

Dear Editor,

Spontaneous (atraumatic) splenic rupture (SSR) may occur even in a normal healthy spleen; however, in most cases, it happens in an enlarged spleen due to serious infections, malignant diseases before and after therapy, metabolic diseases, vascular, and hematological diseases. Generally, it is always a life-threatening condition that requires an immediate diagnosis and splenectomy. To the best of our knowledge, SSR has been rarely described in lymphoma of the marginal zone although these patients always have significant splenomegaly [1, 2, 3]. We present a patient that could be the third case ever reported.

A 57-year-old male with complaints of epigastric pain radiating towards the left hemithorax and towards the left shoulder, which appeared seven days earlier, was admitted at the surgical department. There was no history of trauma. Two years previously, he had been submitted to laparoscopic cholecystectomy, after which he was told about having a moderately enlarged spleen. Being symptom-free, he ignored the information until recently, when pancytopenia and hepatosplenomegaly were discovered during a routine checkup.

At admission, he was pale but hemodynamically stable, with a moderately enlarged abdomen, epigastric tenderness, hepatosplenomegaly, and signs of free fluid within the abdomen. Laboratory data was as follows: hemoglobin 76 g/l, hematocrit 22%, platelets 90 × 10^9/l, white blood cells 1.7 × 10^9/l (differential leukocyte formula: neutrophils 77%, lymphocytes 13%, monocytes 6%, eosinophils 3%, basophils 1%), lactate dehydrogenase 740 U/l (normal range 220–460 U/l), glycaemia 8.4 mol/l, total proteins 53 g/l (albumin 35 g/l), C-reactive protein 112.7 mg/l (normal range 0–5 mg/l). The rest of biochemistry and clotting screen tests were within normal limits. Bone marrow aspirate was normal.

Multidetector computed tomography (MDCT) showed a mild bilateral pleural effusion, enlarged liver (22 cm), enlarged spleen (25 cm) with signs of rupture at the upper pole and several hypodense zones probably caused by infarctions, as well as free fluid of higher density corresponding serohemorrhagic content (Figure 1).

Left subcostal laparotomy was done, through which serohemorrhagic fluid was aspirated, a 4.5-cm rupture of the upper pole of the spleen was found, and splenectomy and biopsy of the liver were performed. The intraabdominal lymph nodes were not enlarged.

The spleen was 280 × 180 × 120 mm in diameter, weighing 2700 g, firm and with a ruptured upper pole. On cross section it showed a large homogenous granular structure. Histology was significant for congestion of the red pulp and proliferation of the white pulp, showing merging and spreading of diffuse infiltrates into the surrounding red pulp. The infiltrate was composed of small and medium sized cells of lymphoid and monocytic morphology corresponding with lymphoproliferation in non-Hodgkin lymphoma (NHL). The area of rupture in the upper pole corresponded with necrosis and fresh infarction. Histology of the liver showed small cell infiltration of portal spaces. Immunohistochemistry showed that the malignant cells were CD20+, CD79a+, CD3-, CD5-, bcl-2 -/+, CD10-, CD23-, cyclin D1-, CD38-, CD138-, kappa-, lambda-, DBA+/-, VD25-, CD123-, CD34-, CD8-, CD30-, CD15-, CD61-, TRAP-, and Ki67 positive in about 10% of cells. Histology and immunohistochemistry were consistent with the diagnosis of splenic marginal zone lymphoma (SMZL). Histology of the liver showed small lymphoid cell infiltration within portal spaces with the same histological and immunophenotypic characteristics as those seen in the spleen. Bone marrow aspirate and histology was free of lymphoma. Virology was negative. The final diagnosis of SMZL with liver involvement was established.

The postoperative recovery was uneventful. High number of platelets of 1020 × 10^9/l required treatment with low-molecular-weight heparin and aspirin. The post-splenectomy prophylactic vaccination was performed.

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Correspondence to:
Nikica GRUBOR
University Clinical Center of Serbia
First Surgical Clinic
Department of Hepatobiliary and Pancreatic Surgery
Dr Koste Todorovića 6
11000 Belgrade, Serbia
gruborn13@gmail.com

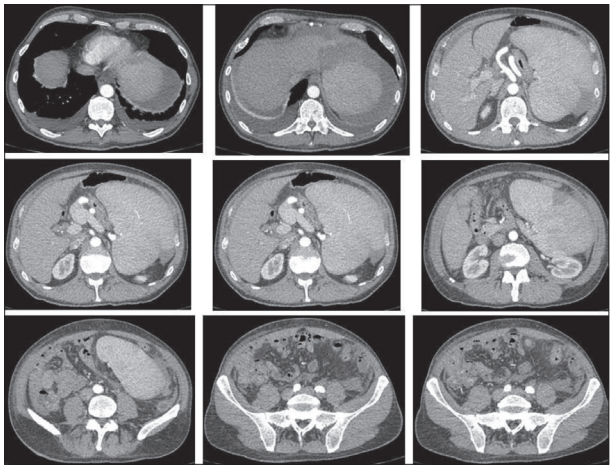

Figure 1. Multidetector computed tomography showing mild pleural effusion, enlarged liver (22 cm), enlarged spleen (25 cm) with signs of rupture in the upper pole and a number of hypodense zones probably caused by infarctions; there is a certain amount of free fluid of higher density corresponding serohemorrhagic content

Although splenectomy has been regarded as a treatment of choice of SMZL, due to liver involvement, additional immunochemotherapy will be applied.

SMZL appears in less than 2% of NHL. Histologically, it shows the proliferation of mature B-lymphocytes. There are three forms of SMZL: nodal MZL, splenic MZL, and extranodal MZL of mucosa-associated tissue. The diagnosis is based on the localization of the disease, histology, and immunohistochemistry. Tumor cells are CD20+, CD79a+, CD5-, CD10-, CD23-, CD43-, and Cyclin D1-. Most frequently they have an indolent course even in patients with bone marrow involvement. If the disease is localized in the spleen only, splenectomy is necessary to establish the diagnosis. In such cases splenectomy is the only treatment requiring no further treatment, but close follow-up is necessary. Rupture of the spleen as in the presented case may be the result of significant enlargement of the spleen due to infiltration of the splenic tissue by the tumorous tissue and infarction in the subcapsular space. Smaller size infarctions may be found in the other areas of the spleen, which may be seen on MDCT, as in our patient.

SSR may have an acute or subacute clinical course, as in the presented case. Thus, clinical pictures of SSR may vary depending on the rupture size, as well as on possible adhesions around the spleen preventing major bleeding in a short period of time. Typically, it is followed by abdominal pain radiating toward the left hemithorax and the left shoulder, as seen in our patient, nausea, vomiting, and hemodynamic changes [4, 5]. In rare cases, pain may be absent, so that possible rupture should be considered if patients with splenomegaly develop a hemodynamic change. In such cases, abdominal ultrasonography and computed tomography are to be performed [6, 7]. In unclear cases, a diagnostic lavage of the peritoneal cavity may be recommended [8]. If the rupture is a major one, serious bleeding can occur and requires urgent diagnosis and surgery; otherwise, it may have a fatal outcome [5, 9, 10]. It is important to know that there is no laboratory or imaging method to predict a rupture, so it is important to take the possibility into account.

Three explanations are offered to clarify the nature of splenic rupture in lymphomas. Parenchyma of the spleen in hematological malignancies becomes infiltrated with malignant cells, which leads to an increase in the intrasplenic pressure, which in turn transmits onto the splenic capsule, causing rupture. The disturbances of coagulation or thrombocytopenia may cause intrasplenic or subcapsular bleeding. Finally, an infarction that leads to rupture may be caused by increased pressure of the proliferated tissue on intrasplenic blood vessels [3, 4, 8, 11].

There are several pathologies that may lead to SSR. In an analysis of 632 publications presenting with SSR, in 845 patients within a period 1980–2008, neoplasms were the cause of SSR in 30.3%, infections (malaria, infectious mononucleosis) in 27.3%, inflammatory non-infectious diseases (autoimmune diseases, hemolytic anemia) in 20%, rupture after different chemotherapeutic agents in 9.2%, mechanical rupture in 6.8%, and idiopathic rupture of the normal spleen in 7% [10, 11].

Several hematological diseases result in splenomegaly. However, SSR in those diseases is an unusual event. Among them, SSR is most frequent in NHL. In a series of 136 published cases of SSR in a period 1861–1995, SSR in NHL was registered in 34%, in acute myeloid leukemia in 34%, and chronic myeloid leukemia in 18% [8, 11]. Among NHL cases, spontaneous rupture appears with similar frequency in diffuse large B-cells lymphomas, blast variant of mantle cell lymphoma, anaplastic large cells, and hepatosplenic lymphoma, and is rarely described in diffuse histiocytic lymphoma, follicular lymphoma, malignant lymphomonocytic, and diffuse histiocytic lymphoma [3, 12]. In this series, no single case of SSR was registered in patients suffering from SMZL lymphoma.

Conflict of interest: None declared.

Nikica Grubor
University Clinical Center of Serbia, First Surgical Clinic
Department of Hepatobiliary and Pancreatic Surgery
Belgrade, Serbia

Nataša Čolović
University of Belgrade
Faculty of Medicine
University Clinical Center of Serbia
Clinic of Hematology
Belgrade, Serbia

Tatjana Terzić
University of Belgrade
Faculty of Medicine
Institute of Pathology
Belgrade, Serbia

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