CASE REPORTS

PRIKAZI SLUČAJEVA

General Hospital Vršac, Department of Surgery, Vršac¹
Institute of Pulmonary Diseases of Vojvodina, Sremska Kamenica
Clinic of Thoracic Surgery²
University of Novi Sad, Faculty of Medicine Novi Sad³

Case report
Prikaz slučaja

Introduction

According to Poulin, enlarged spleen weighing over 1,000 grams is defined as massive splenomegaly, and “severe” if the largest dimension is greater than 20 cm [1]. Splenomegaly is associated with many diseases, but the etiology requires hematological studies on immune response work hypertrophy, red blood cell (RBC) destruction work hypertrophy, congestive (spleenic vein thrombosis or portal hypertension), myeloproliferative, infiltrative (benign or malignant) or purely neoplastic diseases. Differential diagnosis of massive splenomegaly includes several hereditary diseases like Niemann-Pick disease, acid sphingomyelinase deficiency, Gaucher disease, hereditary spherocytosis, thalassemias, and mucopolysaccharidosis. Massive splenomegaly is associated with thrombocytopenia, but also spontaneous infarction and rupture [2]. It is well known that patients with splenomegaly must take additional measures in order to prevent blunt traumas to the abdomen and decrease the risk of splenic rupture [3].

Case Presentation

We present a 46-year-old Caucasian male with a documented medical history of hereditary splenomegaly and thrombocytopenia since childhood. Five years prior to the injury, ultrasound showed the largest diameter of the spleen of 24 cm (Figure 1), and bone marrow aspiration failed to determine the etiology of the disease. However, the tests did reveal pseudo-Gaucher cells and elevated levels of alpha 1 globulin and chitotriosidase, with normal enzyme activity of acid β-glucosidase. The patient was admitted to the surgery department of a rural hospital with symptoms

Summary

Introduction. We present a patient with a delayed rupture of the spleen following a mild abdominal trauma. For years, the patient was treated for hereditary massive splenomegaly with thrombocytopenia, without established etiology. Case Report. The initial non-operative treatment lasted seven days, after which the patient was readmitted to the Emergency Department with signs of intra-abdominal hemorrhage and underwent emergency open splenectomy. Even after pathohistological examination, the etiology of massive splenomegaly remained unknown. Conclusion. Conservative treatment is not recommended in cases of massive splenomegaly; thorough surgical observation in a tertiary care hospital with interventional radiology and a good multidisciplinary team is necessary, while splenectomy is a surgery of choice.

Key words: Splenic Rupture; Splenomegaly; Abdominal Injuries; Delayed Diagnosis; Wounds, Nonpenetrating; Hemorrhage; Splenectomy; Thrombocytopenia; Risk Factors; Diagnosis

Sažetak


Ključne reči: ruptura slezine; splenomegalija; povrede abdomeena; odložena dijagnoza; nepenetrirajuće rane; krvenja; splenektomija; trombocitopenija; faktori rizika; dijagnoza

Corresponding Author: Dr Srđan Putnik, Opšta bolnica Vršac, Hirurško odeljenje, 26300 Vršac. Abraševićeva bb, E-mail: putniksrdjan@outlook.com
of abdominal pain seven days after a mild lower chest trauma. The initial treatment was conservative. The patient was stable; RBC and hematocrit were normal, as well as the blood pressure over the brachial artery, while the ultrasound showed a small amount of fluid above the enlarged spleen (28 x 10 cm) without any intraparenchymal pathological morphology. The patient was discharged and advised to visit a hematologist. Seven days after the initial conservative treatment and two weeks after the trauma, the patient was admitted, this time with hypotension, low hematocrit, and anemia with signs of hemorrhagic shock and increased bilirubin levels. The ultrasound confirmed a large amount of free fluid in the entire peritoneal cavity a large subcapsular hematoma of 20 x 10 cm (Figure 2), and a smaller intraparenchymal hematoma. Emergency laparotomy and splenectomy were performed (Figure 3). No other changes were found during the intervention, especially on the liver. The patient received two units of whole blood. The postoperative period was without complications. The patient was discharged without thrombocytopenia, and received a pneumococcal polyvalent vaccine. The pathology examination revealed a spleen weighing 1,800 grams with several intraparenchymal hematomas from 0.5 to 3 cm, while the largest subcapsular hematoma was 18 x 13 cm (Figure 4). Subsequent histological examination failed to determine the etiology of this massive splenomegaly.

Discussion

Neither the performed splenectomy nor the hematological investigation of our patient with previously known massive splenomegaly succeeded in determining the etiology of the splenomegaly and thrombocytopenia. The patient received no specific therapy for the splenomegaly, apart from being warned not to use nonsteroidal anti-inflammatory drugs. Recent publications do not advise splenectomy for hereditary massive splenomegaly not only due to adverse effects of sepsis but also osteolytic activity [4]. Splenectomy is indicated only in certain circumstances, mainly in patients with severe anemia and life-threatening thrombocytopenia, of which our patient had none. However, he did have a minor trauma for which splenectomy could be indicated [2].

Non-operative management of blunt injuries to the spleen is the treatment of choice for a normal spleen, as well as computerized tomography (CT), provided the patient is hemodynamically stable [5, 6]. In rural hospitals without a CT scanner, the available diagnostic tool is ultrasound, which can also be simply performed as bedside ultrasound. Focused assessment with sonography for trauma (FAST) is indicated in intraperitoneal hemorrhages and intraparenchymal morphology of the spleen [7, 8]. However, special attention must be paid to cases of massive splenomegaly with abdominal pain even after a mild trauma. Upon first admission, our patient had only massive splenomegaly and a small amount of intraperitoneal fluid. He was hemodynamically stable and with normal RBC count. No intraparenchymal or subcapsular hematomas were detected. An attempt was made with conservative treatment. What else can be done for a patient with massive splenomegaly? The first method of choice may include interventional procedure such as splenic artery embolization in a hospital with interventional radiology. The success rate in salvaging the spleen that shows no pathological changes is high, over 90%, but it depends on the severity of injuries [9]. Upon second admission, the ultrasound examination revealed a subcapsular bleeding hematoma, namely grade III, according to the organ injury scale [10]. It is uncertain if splenic artery embolization would be successful in massive splenomegaly and intraparenchymal or subcapsular grade III hematoma. Our hospital has no in-

Figure 1. Splenomegaly five years prior to the injury – ultrasound examination
*Slika 1. Splenomegalija pet godina pre povrede – ultrazvučni pregled

Figure 2. Subcapsular haematoma of the spleen
*Slika 2. Supkapsularni hematom slezine

Abbreviations
RBC – red blood cell
CT – computerized tomography
FAST – focused assessment with sonography for trauma

Figure 1. Splenomegaly five years prior to the injury – ultrasound examination
*Slika 1. Splenomegalija pet godina pre povrede – ultrazvučni pregled

Figure 2. Subcapsular haematoma of the spleen
*Slika 2. Supkapsularni hematom slezine

Abbreviations
RBC – red blood cell
CT – computerized tomography
FAST – focused assessment with sonography for trauma
Interventional radiology or a CT scanner, so the first conclusion is that splenic trauma and massive splenomegaly cases should not be monitored in small hospitals.

Another issue is whether the organ injury scale can be applied in patients with massive splenomegaly. We believe that the standard spleen injury scale is not applicable in cases of massive splenomegaly, whereas non-operative treatment of splenic trauma is not a preferable method [10]. However, each case of a mild trauma in massive splenomegaly should be treated in a hospital not only equipped for conducting interventional and laparoscopic procedures but also has surgeons with experience in splenic resection and a proper hematological support. Delayed rupture of a traumatic hematoma in massive splenomegaly is expected. Upon discharge following splenectomy, our patient had a normal platelet count. The etiology of his disease remains unknown even after histopathological examination.

Clinical follow-up will explain the role of splenectomy in this case of massive splenomegaly of unknown etiology.

Conclusion

Conservative treatment is not recommended in cases of massive splenomegaly; careful surgical observation in a hospital with interventional radiology and a good multidisciplinary team is necessary, while splenectomy is a surgery of choice.

References