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CASE REPORT / ПРИКАЗ БОЛЕСНИКА

Giant spleen as a surgical challenge – case report and literature review

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SUMMARY

Introduction According to the guidelines of the European Association of Endoscopic Surgery, any case where the maximum craniocaudal splenic diameter exceeds 20 cm is considered massive splenomegaly. In addition to metabolic, hematological, and hemodynamic problems, enlarged spleen may cause mechanical difficulties due to the pressure to surrounding organs and vascular structures. The aim of this paper is to present the surgical challenges and technique applied in massive splenomegaly, in a patient who had neglected the importance of regular medical checkups.

Case outline We present a 62-year-old male patient who was admitted to hospital for treatment of previously clinically and radiologically verified splenomegaly but who neglected the importance of regular checkups and medical treatment. Splenectomy was performed with a splenic specimen 38 cm in its maximal diameter.

Conclusion Taking into consideration all the possible benefits and possible complications of surgical treatment, including the quality of life of splenectomized patients, comprehensive preoperative assessment should be made, and surgical treatment selectively applied.

Keywords: splenomegaly; massive splenomegaly; splenectomy

INTRODUCTION

Massive splenomegaly is an enlargement of the spleen that can occur in a wide array of diseases and conditions. According to the guidelines of the European Association of Endoscopic Surgery (EAES), any case where the maximum craniocaudal splenic diameter exceeds 20 cm is considered massive splenomegaly [1].

In addition to metabolic, hematological, and hemodynamic problems, an enlarged spleen may also cause mechanical difficulties. Due to the pressure to surrounding organs and vascular structures, patients often complain of a sense of heaviness in the abdomen, frequent perspiration, nausea, indigestion, etc. [2].

The diagnostics in splenomegaly, depending on the cause, entails a series of procedures, starting with a physical exam, as well as abdominal ultrasonography, computerized tomography (CT), magnetic resonance imaging, laboratory analyses, and finally splenectomy. A definitive diagnosis is established after a histopathological examination [2, 3, 4].

Splenomegaly treatment may be nonsurgical, which includes the treatment of the basic cause of spleen enlargement, as well as surgical. Surgical treatment involves the removal of the spleen, necessitated by the difficulties that its size causes, as well as by rupture or imminent risk of rupture. Also, when preoperative diagnostics does not offer reliable data, splenectomy is performed for diagnostic purposes [5].

The aim of our paper is to present the surgical technique applied in massive splenomegaly, in a patient who had neglected to observe the importance of regular checkups and medical treatment.

CASE REPORT

We present a 62-year-old male patient who was admitted to hospital for treatment of previously clinically and radiologically verified splenomegaly. Abdominal CT detected an enlarged spleen of enormous size – 39.3 cm (Figure 1). At admission, we learned from the patient that he had been diagnosed with an enlarged spleen a number of years before, that he had had bone marrow biopsy several times, and that the results of the biopsies had been normal. We also found out that he had not been keeping up with regular check-ups and examinations during the previous several years. Medical records revealed that he had undergone urinary bladder surgery several years before, due to calculosis, as well as surgery of bilateral inguinal hernia, two years earlier.

Two months before admission to our hospital, the patient visited his doctor complaining of abdominal pain, the sensation of heaviness, discomfort, and increased perspiration. After complete radiological diagnostics had been performed, the patient was examined and tested by a hematologist. Immunophenotyping of

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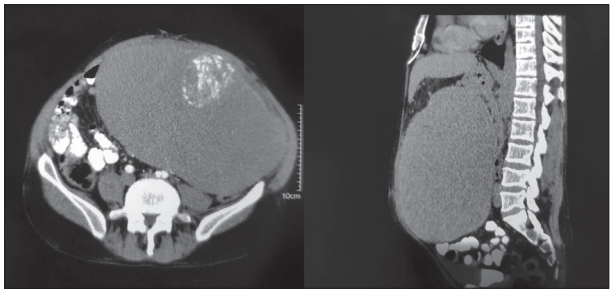

Figure 1. Abdominal computed tomography detected an enlarged spleen of enormous size, 39.5 cm in diameter

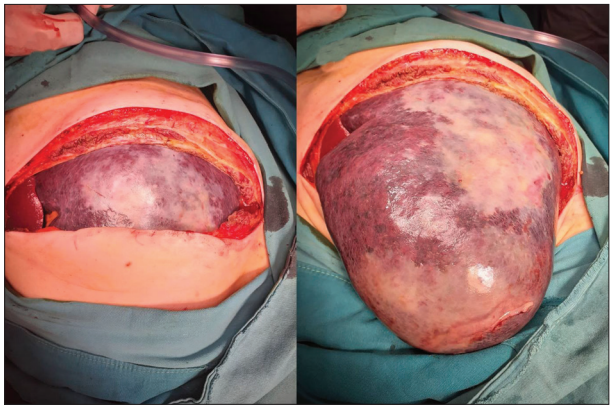

Figure 2. Intraoperative photo shows the enlarged spleen that occupied most of the abdominal cavity

peripheral blood was performed and it was determined that the patient had the B monoclinic population which was Pan B+, CD5-, CD10-, FMC7+ hetero, CD23+ hetero, CD38+ hetero, with IgM/kappa+, with a HILL score of 1. In other laboratory results, there was a marked monoclinic IgM kappa paraprotein in low concentration.

Upon the completion of hematological diagnostics and preparation, further surgical treatment was decided on during a clinical case conference. After preoperative preparations were performed, the patient was placed under general anesthesia and the abdomen was opened with midline laparotomy. The abdomen revealed an enlarged spleen interspersed with white patches (Figure 2), which was pressing the surrounding organs and vascular structures to the opposite side of the abdomen. The next step was the mobilization of the spleen with the aid of the LigaSure device (SurgRx, Redwood City, CA, USA), for severing splenic ligaments. We first identified the lienal

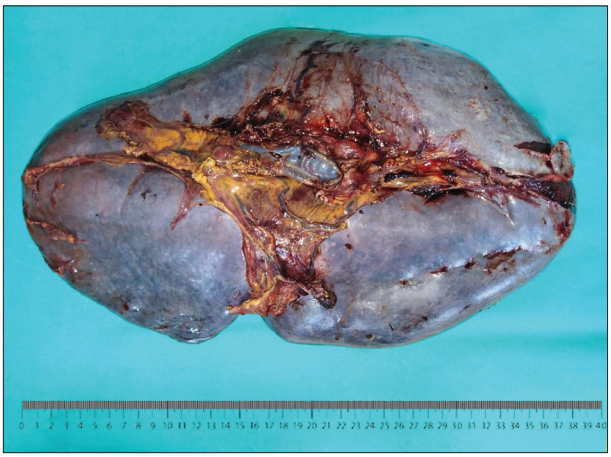

Figure 3. Splenic specimen

artery and vein, which were then, through careful preparation, ligated and severed. Then, we dealt with the short gastric blood vessels, also with the use of LigaSure. After this, splenectomy was completed without incident and the spleen was removed from the abdomen in its entirety. After hemostasis was established, an abdominal drain was placed in the left subphrenic space, and the abdomen was closed by anatomical layers.

The splenic tissue specimen taken after the removal of the spleen from the abdomen (Figure 3) was sent for definitive histopathological examination, which confirmed hairy cell leukemia.

The patient was discharged from hospital on the sixth postoperative day with prescribed antibiotic prophylaxis and a recommendation for post-splenectomy immunization. In addition, the patient was referred for further treatment and follow-up with a hematologist.

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

DISCUSSION

Clinically, massive splenomegaly represents a finding corresponding to a palpable mass in the abdomen below the

Table 1. Causes of splenomegaly

Liver disease	cirrhosis, hepatitis
Hematologic malignancies	lymphomas, leukemias, myeloproliferative disorders
Venous thrombosis	portal or hepatic vein thrombosis
Splenic congestion	venous thrombosis, portal hypertension
Cytopenias	immune thrombocytopenic purpura, autoimmune hemolytic anemia, immune-mediated neutropenia, Felty syndrome
Splenic sequestration	pediatric sickle cell disease, hemolytic anemias, thalassemias
Acute or chronic infection	bacterial endocarditis, infectious mononucleosis, HIV, malaria, tuberculosis, histiocytosis, abscess
Connective tissue diseases	systemic lupus erythematosus, rheumatoid arthritis, adult-onset Still's disease, and some familial autoinflammatory syndromes
Infiltrative disorders	sarcoidosis, amyloidosis, glycogen storage disease
Focal lesions	hemangiomas, abscess, cysts, metastasis

left costal margin at a distance > 8 cm towards the superior iliac spine. The recommendation of the EAES is to use metric units for defining splenomegaly in preoperative imaging diagnostics. The golden standard in diagnosing and determining massive splenomegaly is calculating the splenic volume; however, this procedure requires more time than standard radiological methods [5, 6].

An enlarged spleen can occur within numerous infectious diseases, with hereditary diseases, immunological diseases, as well as in hematological diseases. The most frequent causes of splenomegaly are shown in Table 1 [7].

Pronounced symptoms are mostly present in massive splenic enlargement, which is why patients commonly complain of symptoms manifesting after food intake, of the inability to adequately empty their bladders, of obstructive complaints, as well as of the inability to flex their trunk [8, 9, 10].

In the patient we are presenting, the symptoms were the main reason for his visiting the doctor again. It is our belief, based on insight into the medical records, that bilateral inguinal hernia was directly caused by splenic enlargement and increased pressure within the abdomen.

Splenomegaly can generally be treated with medication, with reductive therapy (radiation therapy), although in massive splenomegaly, potential complications related to this condition should always be taken into consideration – this is why, in this sense, surgical treatment should not be delayed. As a treatment modality in potential rupture, preoperative embolization of the splenic artery should be considered, when there are technical conditions for this procedure, since, in this way, the possibility of bleeding is reduced and partial reduction of the volume of the spleen is achieved, which further facilitates surgical treatment [11, 12, 13].

Since the diagnosis had been established preoperatively and since other treatment options had been exhausted, splenectomy was performed in our patient as a form of palliative treatment, i.e., debulking.

Surgical treatment is a second, sometimes a third therapeutic option, in treating splenomegaly. Taking into consideration all the possible benefits and possible complications of surgical treatment, including the quality of life of splenectomized patients, preoperative assessment should be made, and surgical treatment should be selectively applied, bearing in mind that massive splenomegaly is a great surgical challenge.

Conflict of interest: None declared.

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Масивна спленомегалија као хируршки изазов – приказ болесника и преглед литературе

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САЖЕТАК

Увод Према критеријумима Европског удружења за ендоскопску хирургију масивна спленомегалија подразумева случајеве у којима максимални краниокаудални дијаметар слезине прелази 20 *cm*. Осим метаболичких, хематолошких и хемодинамских поремећаја, значајно увећана слезина може испољавати непожељан компресивни ефекат на суседне органе и васкуларне структуре.

Циљ овог рада је да се прикажу изазови и технике хируршког лечења масивне спленомегалије.

Приказ болесника Болесник стар 62 године са раније дијагностикованом спленомегалијом, који је занемарио значај редовних контролних прегледа, примљен је на клинику због тегоба узрокованих масивном спленомегалијом. После пажљиво спроведене преоперативне припреме одстрањена је слезина краниокаудалног дијаметра 38 *cm*.

Закључак Узимајући у обзир све могуће добробити али и компликације хируршког лечења болесника са масивном спленомегалијом, неопходна је свеобухватна преоперативна хируршка процена и припрема, а хируршко лечење се планира у пажљиво изабраним случајевима.

Кључне речи: спленомегалија; масивна спленомегалија; спленектомија