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Massive splenomegaly due to B-cell lymphoma: A case report

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ABSTRACT

INTRODUCTION: Massive splenomegaly is indicated by spleen weight exceeding 1000 g and largest spleen dimension greater than 20 cm Poulin et al. (1998). In many cases, splenectomy is the treatment of choice for massive splenomegaly because it releases the pressure on adjacent organs and also provides a definitive histopathological diagnosis of the underlying cause Iriyama et al. (2010), Radhakrishnan (2018).
PRESENTATION OF CASE: Herein we present a clinical case of disseminated diffuse large B – cell lymphoma, clinical stage IV, with massive splenomegaly. A 53 – year old man complaining of unintentional major weight loss, palpable abdominal mass in the left hemiabdomen and cervical lymphadenopathy, was admited to Department of abdominal surgery, UMC Ljubljana. Abdominal CT scan showed massive spleen, enlarged retroperitoneal and upper mediastinal lymph nodes and cervical lymphadenopathy. Splenectomy was performed and spleen was sent on histological analysis. Operation and postoperative course were uneventful. Spleen specimen weighed 5034 g (6% of patient body weight) and measured 33 × 24 × 10 cm. Histological and immunohistochemical analysis set the diagnosis of diffuse large B – cell lymphoma. Patients received 5 cycluses of R-CHOP chemotherapy and 2 cycluses of prophylactic intrathecal chemotherapy postoperatively.
DISCUSSION: Splenomegaly in combination with weight loss and malaise is very suggestive of underlying neoplastic condition and therefore requires further diagnostic investigations Han et al. (2008). Splenectomy in combination with adjuvant chemotherapy is the treatment of choice in case of spleen infiltration rith tumorous cells of B-cell lymphoma. However there are other possibilities in diagnosing and treatment of massive splenomegaly, including percutaneous image guided splenic needle biopsy and splenic artery embolisation prior to splenectomy.
CONCLUSION: Our aim with this case report is to present splenectomy in conjuction with chemotherapy as a safe option of treatment for massive splenomegaly due to B-cell lymphoma infiltration.
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1. Introduction

Massive splenomegaly is indicated by spleen weight exceeding 1000 g and largest spleen dimension greater than 20 cm [1]. Diffuse large B-cell lymphoma is the most frequent variant of non-Hodgkin's lymphoma (NHL), representing approximately 33% of all cases [2]. Compared with Hodgkin lymphoma, NHL is much less predictable and has a far greater predilection to disseminate to extranodal sites, including spleen. The prognosis depends on the histologic type, stage, and treatment [3]. The most common malignant cause of massive splenomegaly in developed world is lymphoma. In many cases, splenectomy is the treatment of choice for massive splenomegaly due to lymphoma. Splenectomy not only releases the pressure on adjacent organs and solves the issues with hypersplenism, but it also provides a definitive histopathological diagnosis of the underlying cause [2,4].

Patients after splenectomy are at high risk of infection, primarily by encapsulated bacteria such as Streptococcus pneumoniae, *Haemophilus influenzae* type B, and Neisseria meningitides. Prevention of postsplenectomy infection depends on education of patients, adoption of appropriate vaccination schedules, and use of prophylactic antibiotics [5].

The work has been reported in line with the SCARE criteria [6]. Informed consent was obtained in this study.

2. Case report

A 53- year old, previously healthy man, presented with malaise, weight loss of 30 kg in 8 months, mild dyspnea and cervical lymphadenopathy. He was a former smoker. Other than that, his medical history was unremarkable. On physical examination, he had a palpable massive spleen reaching 4 cm under spina iliaca anterior superior and crossing the midline 8 cm to the right side. He also had mild hepatomegaly with no signs of free fluid in abdominal

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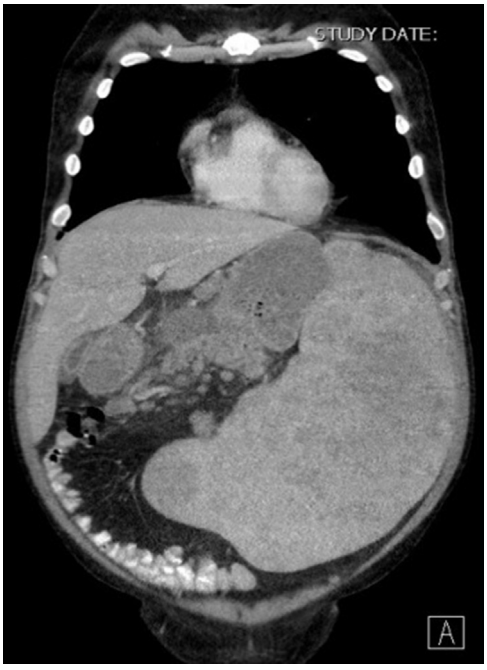

Fig. 1. Computed tomography showing massive splenomegaly.

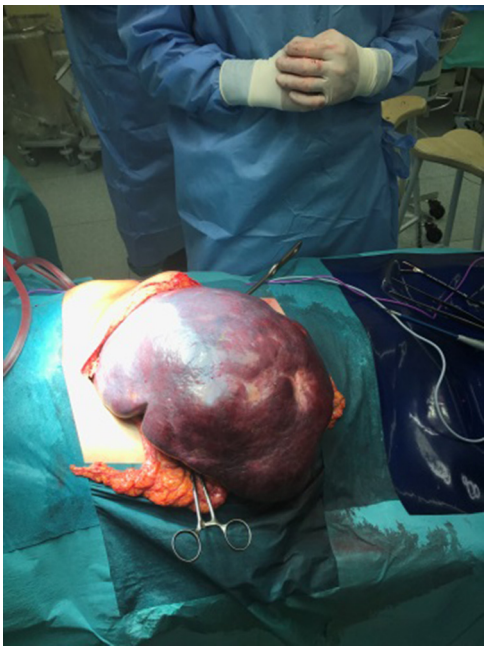

Fig. 2. Intraoperative photo showing extent of splenomegaly.

cavity. On right side of neck, there was 10 x 8 cm large solid lymph node aggregate. He also had mild pleural effusion.

Laboratoy results showed pancytopenia and elevated level of LDH (10,8 mmol/L). CT scan revealed a massive spleen (Fig. 1), moderately enlarged liver, enlarged retroperitoneal and upper mediastinal lymph nodes. Aspiration biopsy of cervical lymph nodes was performed and set the diagnosis of B-cell lymphoma. Patient was advised to proceede with splenectomy in order to prevent possible trombosis of abdominal veins due to compression of massive spleen. At first, patient rejected the operative treatment, but changed his mind later. Preoperatively he was vaccinated with Pneumovax, *Haemophilus* B and meningococcal vaccine.

On the day of operation patient was placed in right lateral decubitus position. Upper midline incision was made and extended in left subcostal incision in order to gain access. Mobilisation of spleen began with dissection of splenocolic, phrenicosplenic, gastrosplenic ligaments and short gastric arteries. Splenic artery was covered with fibrous material and ended in splenic hylum which was also fibrotic and thickened. Splenic vessels and hylum were divided with 60 mm vascular stapler. After that spleen was removed from abdomen (Fig. 2), CH 21 abdominal drain was inserted in the left upper quadrant. The resected spleen weighed 5034 g, measured 33 × 24 x 10 cm and comprised 6% of patient body weight (preoperatively 81 kg).

Upon histological analysis, spleen was completely infiltrated with confluating nodes of whitish grey tissue. Immunohistochemical staining confirmed the diagnosis of diffuse large B-cell lymphoma. First postoperative day patient started receiving antibiotic prophylaxis with Penicillin 1000 mg/12 h which will be continued for two years. On fifth postoperative day patient was transferred to oncology department, where he received 5 cycluses of R-CHOP chemotherapy and 2 cycluses of prophylactic intrathecal chemotherapy in the following months respectively.

3. Discussion

We reported a case of diffuse large B – cell lymphoma presenting with massive splenomegaly and hypersplenism. Splenomegaly in combination with weight loss and malaise is very suggestive of underlying neoplastic condition and therefore requires further diagnostic investigations [7]. Our patient had large solid lymph node aggregate on the right side of his neck as well, which appeared to be convenient for needle biopsy diagnosis. The results of biopsy pointed to B - cell lymphoma. CT scan showed massive spleen with enlarged lymph nodes in thorax and abdomen in the context of systemic disease.

Splenectomy was therefore performed in order to relieve the symptoms, reduce the hypersplenism, prevent spleen rupture and confirm the diagnosis of lymphoma. However, there are several reports in the literature, discussing the risk of splenectomy for massive (greater than 1500 g) splenomegaly. Perioperative mortality of mentioned procedure is high at about 20%. In some cases with poor general status, bleeding tendency, complications due to infection or organ failure, one should be hesitant to use an invasive diagnostic method. On the other hand, splenic needle biopsy may provide an adequate diagnosis without severe complications. Tam et al reported percutaneous image guided splenic needle biopsy in 156 consecutive cases and concluded that splenic needle biopsy in the evaluation of new or recurrent neoplasm is a minimally invasive procedure with low complication rates and a high diagnostic yield. Therefore, if institutionally and technically possible, splenic needle biopsy should be taken into consideration for high risk patients [10].

Another interesting technique of massive splenomegaly operative treatment is splenic artery embolisation (SAE) prior to splenectomy. SAE is achieved with microcoils or with intraluminal balloon embolization and subsequent continuous epinephrine infusion for reducing the spleen volume. It enables easier handling of the spleen and minimizes bleeding and is especially appropriate for laparoscopic splenectomy of massive splenomegaly [11].

Most cases of aggressive lymphoma such as diffuse large B – cell lymphoma show fast disease expansion and progression, requiring immediate chemotherapy. It is speculated that the high rate of perioperative mortality in massive splenomegaly could be due to rapid progression of disease and such patients should be subjected to less invasive diagnostic methods and treated immediately [12]. Perioperative and postoperative course with our patient progressed without complications, on the other hand.

Splenomegaly underlying lymphoma is often reported as a cause of hypersplenism. After splenectomy, cytopenias resolve in most cases. This was also the case with our patient in the weeks after splenectomy [8]. That being said, splenectomy is useful not only for diagnosis but also for treatment of underlying hematologic malignancy.

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of this article.

Conflicts of interest

There are no conflicts of interest.

Sources of funding

There were no sponsors participating in this case report.

Ethical approval

Our institution does not require ethical approval for publishing a case report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Author contribution

Study conception and design: Mihajlo Djokic, M.D.
Acquisition of data: Bostjan Plesnik, M.D.
Analysis and interpretation of data: Bostjan Plesnik, M.D.
Drafting of manuscript: Miha Petric, M.D.
Critical revision: assist. prof. Blaz Trotovsek, M.D.

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Guarantor

Mihajlo Djokic, M.D., Bostjan Plesnik, M.D.

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