Case report

Primary splenic lymphoma discovered on massive splenomegaly: A case report

Hazem Beji a,b,*,**,***, Mahdi Bouassida a,b, Ghazi Laamiri a,b, Emna Chelbi c,b, Salwa Nechi c,b, Hassen Touinsi a,b

a Department of General Surgery, Hospital Mohamed Taher Maamouri, Nabeul, Tunisia
b University Tunis El Manar, Faculty of Medicine of Tunis, Tunisia
c Department of Pathology, Hospital Mohamed Taher Maamouri, Nabeul, Tunisia

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ABSTRACT

Introduction and importance: Malignant lymphoma occurs in all the systemic organs. Rarely, large B-cell lymphoma is located in the spleen, making the diagnosis difficult. Herein, we report a patient presenting with massive splenomegaly due to LBCL. Splenectomy was essential to assess the diagnosis and to guide postoperative therapeutics.

Presentation of a case: A 47-year-old woman, with no comorbidities, complained of weight loss and abdominal pain. She had a palpable spleen that extended below the navel. CT scan revealed massive splenomegaly and lymph nodes in the spleen hilum. Splenectomy was performed. Histopathological examination confirmed the diagnosis of large B-cell lymphoma. The postoperative course was uneventful. Three courses of chemotherapy were given. The patient was in remission after a follow-up of 8 months.

Discussion: Massive splenomegaly can be one of the circumstances of the discovery of large B-cell lymphoma. Splenectomy was then essential to confirm the diagnosis and to guide postoperative therapeutics. It also permits reducing hypersplenism and preventing spleen rupture. In patients with high operative risk, splenic needle biopsy should be taken into consideration. Splenic artery embolization before surgery can also be performed in patients having massive splenomegaly to reduce the spleen volume.

We highlight the importance of splenectomy to confirm the diagnosis and to relieve the symptoms. Postoperative chemotherapy is essential to prevent relapses.

Conclusion: Splenectomy is essential in spleen localized large B-cell lymphoma. It permits to confirm the diagnosis, relieve symptoms, and treatment of underlying hematologic malignancies. Postoperative chemotherapy is essential to prevent relapses.

1. Introduction and importance

Malignant lymphoma occurs in all the systemic organs. In comparison with Hodgkin Lymphoma, Non-Hodgkin's Lymphoma (NHL) has a greater predilection to touch extranodal sites, including the spleen [1,2]. Rarely, large B-cell lymphoma is located only in the spleen, making the diagnosis difficult. Primary splenic lymphoma is a very rare entity. It is a disease confined to the spleen or at the most involves hilar lymph nodes with no recurrence after splenectomy [3,4]. The place of splenectomy is still debated [5]. It is mainly important in providing histopathological confirmation, relieving the symptoms, and reducing hypersplenism.

Herein, we report a patient presenting with massive splenomegaly due to spleen localized LBCL. Splenectomy was essential to assess the diagnosis. This work has been reported in line with the SCARE 2020 criteria [6].

* Corresponding author at: 52 - Street of roses, 8050 Hammamet, Tunisia.
** Corresponding author at: University of Tunis El Manar, Faculty of Medicine of Tunis, Tunisia.
*** Corresponding author at: Department of General Surgery, Hospital Mohamed Taher Maamouri, Nabeul, Tunisia.
E-mail address: Hazembj@gmail.com (H. Beji).
2. Presentation of a case

A 47-year-old woman, with no comorbidities, complained of weight loss and abdominal pain.

On the physical examination, she had no palpatible peripheral lymphadenopathy. She had a palpable spleen that extended below the navel. There were no ascites or hepatomegaly.

Laboratory studies revealed haemoglobin of 9.3 g/dl, platelet count of 75,000/μl, white blood cells count of 5300/μl. Renal and hepatic functions were normal. LDH, uric acid, and serum Ca levels were normal.

CT scan revealed massive splenomegaly and lymph nodes in the spleen hilum (Figs. 1 and 2). Splenectomy was planned.

The two essential diagnoses evoked were: lymphoma and abdominal tuberculosis.

We opted for laparotomy instead of laparoscopy because of the massive splenomegaly. A left subcostal incision was made. Intraoperative findings revealed lymph nodes in the splenic hilum. We performed splenectomy. It was performed by a surgeon with seven-year experience. The blood loss was estimated at 150 mL and the operation time was 150 min.

The resected spleen weighed 3.5 kg and measured 22 × 20 × 10 cm.

In the cut surface, there were grey-white homogenous tumors soft in consistency (Fig. 3).

Microscopy showed the proliferation of large neoplastic lymphoid cells (Fig. 4). Immunohistochemical staining confirmed the diagnosis of large B-cell lymphoma. The tumor cells were immunopositive for CD 20 (Fig. 5).

The disease was classified low risk according to the international prognostic index for non-Hodgkin lymphoma [7].

Fig. 1. Sagittal CT scan showing massive splenomegaly and splenic hypo-dense lesions.

Fig. 2. Coronal CT scan showing massive splenomegaly and lymph nodes in the splenic hilum.
adhesions to surrounding structures, challenges with retraction and retrieval, and possible trauma to dilated veins or the splenic capsule with subsequent bleeding, laparotomy is the gold standard treatment according to the European Association for Endoscopic Surgery (EAES) [13].

Splenectomy was then essential to confirm the diagnosis and to guide postoperative therapeutics. It also permits reducing hypersplenism and preventing spleen rupture [1]. Splenic artery embolization before surgery can also be performed in patients having massive splenomegaly to reduce the spleen volume. It reduces bleeding and makes spleen manipulation simpler [14].

It was reported that splenectomy for massive splenomegaly has a high rate of perioperative mortality, nearing 20% [15,16]. This could be due to the rapid progression of the disease [17].

Patients after splenectomy are at significant risk of infection by encapsulated bacteria such as Streptococcus pneumoniae, Haemophilus influenzae type B, and Neisseria meningitidis. Prophylactic antibiotics along with the adoption of appropriate vaccination are necessary to prevent post-splenectomy infections [18]. Postoperative anticoagulation therapy is mandatory to avoid portal or splenic vein thrombosis [9].

Most cases of aggressive lymphoma such as large B-cell lymphoma show fast disease expansion and progression, requiring immediate adjuvant chemotherapy [1].

In summary, we reported a case of spleen localized lymphoma in a patient presenting massive splenomegaly and hypersplenism. We highlight the importance of splenectomy to confirm the diagnosis, relieve the symptoms, reduce hypersplenism, and avoid splenic rupture.

4. Conclusion

Splenectomy is essential in spleen localized large B-cell lymphoma. It permits to confirm the diagnosis, relieve symptoms, and treatment of underlying hematologic malignancies. Postoperative chemotherapy is essential to prevent relapses.

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Ethical approval

Not required.

Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Guarantor

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Dr. Mahdi Bouassida

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CRediT authorship contribution statement
Hazem Beji and Mahdi Bouassida did the conception and design of the work, the data collection, and the data analysis and interpretation. Ghazi Laamiri and Emna Chelbi did the critical revision of the article.

Declaration of competing interest
No conflicts of interest.

References
[1] M. Djokic, B. Plesnik, M. Petric, B. Trotovsek, Massive splenomegaly due to B-cell lymphoma: a case report, Int. J. Surg. Case Rep. 48 (2018) 76–78, https://doi.org/10.1016/j.ijscr.2018.05.013. Epub 2018 May 29. PMID: 29885916; PMCID: PMC6041427.
[2] S. Le Gouill, Mantle cell lymphoma: an overview from diagnosis to future therapies, La Revue de Medecine Interne. 31 (9) (2010 Sep) 615–620, https://doi.org/10.1016/j.revmed.2009.05.016. PMID: 20488592.
[3] T. Das Gupta, B. Goombes, R.D. Brosfeld, Primary malignant neoplasms of the spleen, Surg. Gynecol. Obstet. 120 (1969) 947–960.
[4] L. Cavanna, F. Artioli, D. Vaillanu, C. Di Donato, R. Bertè, C. Carapezzi, R. Foroni, C. Del Vecchio, B. Lo Monaco, R. Prati, Primary lymphoma of the spleen. Report of a case with diagnosis by fine-needle guided biopsy, Haematologica 80 (1995) 241–243 [PMID: 7672717].
[5] M. Mollejo, P. Algara, M.S. Mateo, J. Menarguez, E. Pascual, M.F. Fresno, F.I. Camacho, M.A. Pirs, Large B-cell lymphoma presenting in the spleen: identification of different clinicopathologic conditions, Am. J. Surg. Pathol. 27 (7) (2003 Jul) 895–902, https://doi.org/10.1097/00000478-200307000-00004. PMID: 12826881.
[6] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, for the SCARE Group, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (2020) 226–230.
[7] The international non-Hodgkin's lymphoma prognostic factors project. A predictive model for aggressive non-Hodgkin's lymphoma, N. Engl. J. Med. 329 (14) (1993 Sep 30) 987–994. PubMed ID: 8141877.
[8] N. Iriyama, A. Horikoshi, Y. Hatta, Y. Kobayashi, S. Sawada, J. Takeuchi, Localized, splenic diffuse large B-cell lymphoma presenting with hypersplenism: risk and benefit of splenectomy, Intern. Med. 49 (2010) 1027–1030.
[9] S.B. Ingle, C.R. Ingle, Splenic lymphoma with massive splenomegaly: case report with review of literature, World J. Clin. Cases 2 (9) (2014 Sep 16) 478–481, https://doi.org/10.12998/wjcc.v2.i9478. PMID: 25232555; PMCID: PMC4163774.
[10] A. Tam, S. Krishnamurthy, E.P. Pillisbary, J.E. Emsro, S. Gupta, R. Murthy, K. Ahrr, M.J. Wallace, M.E. Hicks, D.C. Madoff, Percutaneous image-guided splenic biopsy in the oncology patient: an audit of 156 consecutive cases, J. Vac. Interv. Radiol. 19 (2008) 80–87, https://doi.org/10.1016/j.jvir.2007.08.025. PMID: 18192471.
[11] A. Guirat, M. Koubba, R. Mzali, B. Abid, S. Ellouz, N. Affes, M. Ben Jemaa, F. Frikha, M. Ben Amar, M.I. Beyrouti, Peritoneal tuberculosis, Clin. Res. Hepatol. Gastroenterol. 35 (1) (2011 Jan) 60–69, https://doi.org/10.1016/j.cgh.2010.07.023. PMID: 21215540.
[12] V.P. Koshekov, Z.H. Nemeth, M.S. Carter, Laparoscopic splenectomy: outcome and efficacy for massive and supramassive spleens, Am. J. Surg. 203 (2012) 517–522.
[13] B. Habermalz, S. Sauerland, G. Deckcr, et al., Laparoscopic splenectomy: the clinical practice guidelines of the European Association for Endoscopic Surgery (EAES), Surg. Endosc. 22 (2008) 821–848.
[14] T. Nitta, K. Fujii, H. Kawasaki, I. Taksaka, S. Kawata, M. Onaka, T. Ishibashia, Efficacy and surgical procedures of preoperative splenic artery embolization for laparoscopic splenectomy of a massive splenomegaly: a case report, Int. J. Surg. Case Rep. 16 (2015) 174–176.
[15] B. Han, Z. Yang, T. Yang, W. Gao, X. Sang, Y. Zhao, T. Shen, Diagnostic splenectomy in patients with fever of unknown origin and splenomegaly, Acta Haematol. 119 (2008) 83–88, https://doi.org/10.1159/0001186327. PMID: 18305380.
[16] D.N. Danforth, D.L. Fraker, Splenectomy for the massively enlarged spleen, Am. Surg. 57 (1991) 108–113 [PMID: 1992865].
[17] B. Pottakkat, R. Kashyap, A. Kumar, S.S. Sikora, R. Saxena, V.K. Kapoor, Redefining the role of splenectomy in patients with idiopathic splenomegaly, ANZ J. Surg. 76 (2006) 679–682.
[18] A.D. Jones, M. Khan, J. Cheshire, D. Bowley, Postsplenectomy prophylaxis: a persistent failure to meet standard? Open forum, Infect. Dis. 3 (2016), ofw197 (October (4)).