a whole-body PET scan and BM trephine biopsy.

In conclusion, we describe a case of an elderly and frail patient with primary DLBCL of the BM that was discovered during work-up for pancytopenia, which was initially suspected to be caused by MDS; the patient achieved long-lasting CR that was maintained for 20 months following the completion of eight courses of R-CHOP. Primary DLBCL of the BM is a rare but distinctive form of extranodal NHL with a poor prognosis and a reported 2-year survival rate of approximately 30% and median survival time of 14.9 months [4]. Given its rarity, no standard therapy for primary DLBCL of the BM has been established. However, as R-CHOP has become the standard treatment for DLBCL in elderly patients, this case suggests that, similar to other types of DLBCL, this regimen can be a well-tolerated, front-line treatment with an efficacy to allow durable CR, even in DLBCL cases with poor prognosis such as ours.

The follow-up period for this patient was too short to draw any firm conclusions; however, compared with the usually dismal outcome of this type of NHL, the 24-month continuous clinical response observed in this case would be considered a satisfactory therapeutic result. In this context, R-CHOP can be considered a reasonable approach in cases where advanced age, toxicity concerns, and comorbidities may detract from the pursuit of more aggressive treatments such as high-dose chemotherapy and ASCT.

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REFERENCES

1. Martinez A, Ponzoni M, Agostinelli C, et al. Primary bone marrow lymphoma: an uncommon extranodal presentation of aggressive non-hodgkin lymphomas. Am J Surg Pathol 2012;36:296-304.
2. Gudgin E, Rashbass J, Pulford KJ, Erber WN. Primary and isolated anaplastic large cell lymphoma of the bone marrow. Leuk Lymphoma 2005;46:461-3.
3. Chang H, Hung YS, Lin TL, et al. Primary bone marrow diffuse large B cell lymphoma: a case series and review. Ann Hematol 2011;90:791-6.
4. Strauchen JA. Primary bone marrow B-cell lymphoma: report of four cases. Mt Sinai J Med 2003;70:133-8.
5. Kajiura D, Yamashita Y, Mori N. Diffuse large B-cell lymphoma initially manifesting in the bone marrow. Am J Clin Pathol 2007;127:762-9.
6. Hishizawa M, Okamoto K, Chonabayashi K, Kaneko H, Watanabe M, Tsudo M. Primary large B-cell lymphoma of the bone marrow. Br J Haematol 2007;136:351.
7. Matthies A, Schuster SJ, Alavi A. Staging and monitoring response to treatment in primary non-Hodgkin's lymphoma of bone marrow using (18)F-fluorodeoxyglucose positron emission tomography. Clin Lymphoma 2001;1:303-6.
8. Kazama H, Teramura M, Yoshinaga K, Masuda A, Motoji T. Long-term remission of primary bone marrow diffuse large B-cell lymphoma treated with high-dose chemotherapy rescued by in vivo rituximab-purged autologous stem cells. Case Rep Med 2012;2012:957063.

Late but effective long-term response to splenectomy in the treatment of immune thrombocytopenia

TO THE EDITOR: Splenectomy is the preferred treatment for steroid-refractory immune thrombocytopenia (ITP) [1]. The initial response to splenectomy is rapid and characterized, in most cases, by immediate thrombocytosis; this finding is considered a favorable predictive factor for long-term response to splenectomy in ITP patients [2, 3]. If platelet count does not increase after splenectomy, it may indicate failure of the procedure and lead providers to consider other salvage treatments.

We report a case of a late response to splenectomy in a patient who was diagnosed with ITP at another center in 1991 at the age of 23 years. At primary diagnosis, a comprehensive work-up for underlying disorders was unremarkable, except for slight positivity for antinuclear antibodies (ANA). The patient was initially prescribed prednisone at a dosage of 1 mg/kg body weight/day; after approximately 1 year, the dosage was appropriately tapered and administration of the steroid was eventually stopped. However, shortly thereafter, the patient experienced a relapse of ITP. She refused splenectomy; therefore, prednisone was restarted and continued at the lowest effective dosage (25–30 mg/day) that allowed stable maintenance of a platelet count $>30\times10^9$/L until 2006. During this time, she was followed up at different hematologic centers. In 2006, the patient was reevaluated because of unresponsiveness to prednisone and was found to have heterozygous factor V Leiden mutation, with no other abnormalities. She refused splenec-

tomy once again. Therefore, cyclosporin A was administered as second-line treatment at a dosage of 200 mg/day, and a stable increase in the platelet count ($>50\times10^9$/L) was achieved.

In December 2010, the patient developed hypertension for which she received antihypertensive therapy and presented to our hospital in June 2011. A comprehensive work-up revealed hypertension and mild renal failure that were related to cyclosporin A. Other underlying or associated medical conditions were carefully excluded. In particular, comprehensive laboratory panel results for autoimmunity, including ANA, were negative; serological test results for HIV and hepatitis viruses were also negative. Therefore, cyclosporin A was discontinued and prednisone was restarted. Following the hypertensive crisis, the patient was admitted with a platelet count of 15×10^9/L and intravenous immunoglobulin (IVIG) was administered. In November 2011 (21 years after the primary ITP diagnosis), at the age of 44 years, the patient underwent laparoscopic splenectomy with IVIG and enoxaparin prophylaxis. After the procedure, the patient developed pneumonia and acute pulmonary embolism and was admitted to the intensive care unit (ICU); however, after laparoscopic splenectomy, the platelet count did not increase and additional IVIG was required. The patient was discharged from the ICU on full enoxaparin alone for 6 months. Following laparoscopic splenectomy, the platelet count spontaneously increased to $>50\times10^9$/L and $>100\times10^9$/L after 2 and 7 months, respectively. The patient is well and active approximately 2 years and 22 years after laparoscopic splenectomy and ITP diagnosis, respectively; her most recent platelet count was 550×10^9/L, for which she is receiving low-dose acetylsalicylic acid.

Our case outlines the effective role of splenectomy in advanced and long-term ITP as well as the possible occurrence of a late response to the procedure. Adult ITP patients who experience a relapse after initial corticosteroid treatment present a therapeutic challenge [1]. The current guidelines recommend splenectomy in this setting; indeed, it results in a high rate of durable responses and the possibility of a definitive cure in a remarkable proportion of patients [1]. However, ITP patients may be at increased risk of perioperative and long-term complications, such as venous thromboembolism and sepsis [4]. These concerns should be considered in the selection of newer available treatments that could potentially allow deferment of splenectomy but do not offer complete amelioration [5].

In the present case, the patient maintained a sustained response 2 years after splenectomy, although the follow-up period was not sufficient to consider her definitively cured. As in our case, the decision to proceed to splenectomy should be patient-specific and performed after the patient has been adequately informed of the potential risks and benefits of the procedure compared with those of alternative treatment options, including the newer thrombopoietin-receptor agonists that require continuous administration to effectively maintain platelet count responses without possibility of complete amelioration.

In conclusion, we report a specific post-splenectomy outcome, characterized by a slow and delayed increase in platelet count, in a patient with long-term, advanced, and steroid-refractory ITP who could not continue treatment with cyclosporin A because of the significant toxicity induced by this agent. The platelet count after splenectomy is considered an important prognostic factor; in particular, a high platelet count on day 7 and in the third month has been reported as predictors of long-term response [2, 3]. Our patient experienced a very slow increase in platelet count; indeed, following laparoscopic splenectomy, the platelet count spontaneously increased to $>50\times10^9$/L and $>100\times10^9$/L after 2 and 7 months, respectively. This particular platelet kinetics have not been fully understood and are attributed to a potential detrimental effect on megakaryocytopoiesis by the medical complications experienced by our patient, such as pulmonary embolism and infection.

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REFERENCES

1. Provan D, Stasi R, Newland AC, et al. International consensus report on the investigation and management of primary immune thrombocytopenia. Blood 2010;115:168-86.
2. Ojima H, Kato T, Araki K, et al. Factors predicting long-term responses to splenectomy in patients with idiopathic thrombocytopenic purpura. World J Surg 2006;30:553-9.
3. Wang M, Zhang M, Zhou J, et al. Predictive factors associated with long-term effects of laparoscopic splenectomy for chronic immune thrombocytopenia. Int J Hematol 2013;97:610-6.
4. Boyle S, White RH, Brunson A, Wun T. Splenectomy and the incidence of venous thromboembolism and sepsis in patients with immune thrombocytopenia. Blood 2013;121:4782-90.
5. Stasi R, Newland A, Thornton P, Pabinger I. Should medical treatment options be exhausted before splenectomy is performed in adult ITP patients? A debate. Ann Hematol 2010;89:1185-95.