Revista Brasileira de Hematologia e Hemoterapia
Brazilian Journal of Hematology and Hemotherapy
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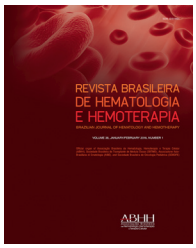

Scientific Comment

Diffuse large B cell gastric lymphoma a rare disease: the effort to obtain scientific data in a multicenter, multinational retrospective trial☆

*Nelson Hamerschlak**

Hospital Israelita Albert Einstein, São Paulo, SP, Brazil

Primary diffuse large B cell gastric lymphoma (DLBCGL) accounts for approximately 1.5% of gastric neoplasms and 5% of lymphomas.[1] It represents 30–40% of the gastrointestinal lymphomas and it is the most common extranodal site of lymphoma. Generally, it occurs in patients between 50 and 60 years old and there is a slight male predominance.[2]

The most common symptom is epigastric pain but patients can present with anorexia, nausea or vomiting, weight loss and bleeding. Endoscopy and biopsy should be performed in this kind of patient for the correct diagnosis. Some times pinch biopsies may miss the diagnosis and larger samples have to be attained.[3–5] The use of endoscopic ultrasound and flow cytometry may achieve higher accuracy rates.[2–5]

An association of *Helicobacter pylori* with DLBCGL exists and treatment to eradicate *H. pylori* has to be used in the management of the disease when necessary.[6] Very few patients outside clinical trials are candidates for only *H. pylori* therapy.

The Ann Arbor staging system is not always considered as a tool in gastrointestinal lymphomas but there is limited consensus in the use of several other staging systems.[7]

Treatment options for DLBCGL include surgery, radiation therapy and chemoimmunotherapy with or without treatment for *H. pylori* or a combination of the above. Surgery is used in patients with complications such as perforation, severe bleeding or obstruction. Nowadays, the majority of the patients are treated with rituximab-based chemotherapy with or without radiotherapy consolidation.[8,9]

The paper by Delamain et al., published in this issue of the Revista Brasileira de Hematologia e Hemoterapia (RBHH), addresses the main findings of DLBCGL in a multicenter retrospective study with 104 patients from three countries, Brazil, Portugal and Italy. This kind of study is very important in rare diseases since the majority of the recommendations are based on data from case series, rather than large randomized trials.[10]

Different to the literature, the authors found a slightly higher incidence in women, advanced stage and high-risk age-adjusted international prognostic index (aaIPI) at diagnosis. The median age was in agreement with the current literature and 13% of the patients were positive for *H. pylori*. Few patients required surgery because of complications. The patients were treated with rituximab with the cyclophosphamide, doxorubicin hydrochloride, vincristine sulfate and prednisone regimen (R-CHOP) with or without radiotherapy and the authors got impressive results considering not only complete remission, but also overall survival rates. They also contribute to the literature by identifying the aaIPI as a predictor factor for the survival of patients.

DOI of original article: http://dx.doi.org/10.1016/j.bjhh.2016.04.010.

☆ See paper by Delamain et al. on pages 247–51.

* *Corresponding author at*: Centro de Pesquisa Clínica, Instituto Israelita de Ensino e Pesquisa Albert Einstein, Av. Albert Einstein, 627/520, 05651-901 São Paulo, SP, Brazil.

E-mail address: hamer@einstein.br

http://dx.doi.org/10.1016/j.bjhh.2016.05.014

Conflicts of interest

The author declares no conflicts of interest.

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