scopy and gastric biopsy was normal. No viral infection was detected. The molecular study of the PB lymphocytes by polymerase chain reaction detected a monoclonal pattern.

Corticosteroid therapy was started with prednisone at 120 mg/day dose. After 2 weeks of treatment, no improvement was observed. Chlorambucil was initiated at dose of 12 mg/day, reaching normal values (2.58×10^9/L lymphocytes) after 7 days of chlorambucil therapy. Flow cytometry analysis showed disappearance of the previous clonal population. Nevertheless, the patient experienced a progressive worsening of her neurological symptoms.

Our case was a MCL in leukemic phase. MCL has been proposed as a new specific clinicopathologic entity.[1] Leukemic manifestation of this lymphoma seems fairly uncommon, perhaps because some of these cases are misdiagnosed as chronic lymphocytic leukemia (CLL).[2,3] Some authors postulate that leukemic phase of MCL is indicative of aggressive disease and predicts poor prognosis, although, in some cases leukemic MCL can be an indolent condition and was classified as atypical CLL. The prognosis does not appear as ominous in those patients in whom a leukemic phase appears at diagnosis, as compared with development during the clinical course.[4] In our patient, leukemic phase of MCL was identified at diagnosis, but the course appeared to be indolent, since it was well controlled with therapy. The association of combined motor neuron diseases and lymphoma has been recognized[5-7] and a common origin has also been hypothesized.[8]

Acknowledgments

The authors wish to thank Dr. E. Roldán for the cytometry analysis.

Key words

Lymphocytic lymphoma of intermediate differentiation, mantle cell lymphoma, motor neuron disease, amyotrophic lateral disease.

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Pathologic rupture of the spleen as the initial manifestation of acute lymphoblastic leukemia: an additional case

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Pathological rupture of the spleen (PRS) is a rare, but well known complication of some hematological malignancies. In a recent review of the literature, Giagounidis *et al.*[1] identified 136 cases of pathologic rupture of the spleen since 1861. This number gives an idea of how seldom it occurs. In this review, 34% of the cases had occurred in acute leukemias, and 13% in acute lymphoblastic leukemias. In most cases, PRS occurs on the course of the disease. PRS as initial manifestation of ALL is a very rare feature: only six cases have been reported in the literature.[2-6] Before now, in our hospital, we had only seen one case of a patient suffering from chronic myelomonocytic leukemia who presented splenic rupture as the initial manifestation of this disease.[7] We now describe the seventh case, to our knowledge, of acute lymphoblastic leukemia presenting as pathologic splenic rupture.

A 47-year-old man was admitted to our hospital in April of 1997 complaining of sudden onset of pain in the left upper abdomen, which radiated to the left shoulder (Kehr's sign). The patient had been well until the previous week of admission, when he began to have fever, malaise, sweats and fatigue. No previous trauma had occurred. On admission, his blood pressure was 110/80 mmHg and there was tachycardia of 110/min. His abdomen was tender and widely distended, and there were nausea and vomiting. Two nodes, 1.5 cm in diameter, were found in the left supraclavicular region. Laboratory analyses showed a leukocyte count of 14.3×10^9/L with 34% neutrophils, 60% lymphocytes,

5% monocytes, 0.3% eosinophils; hemoglobin was 8.1 g/dL and platelet count 190×10^9/L. Lactate dehydrogenase was 1978 U/L (range, 80-480 U/L). Coagulation tests were normal. The ultrasonographics findings were a spleen of 19 cm of longitudinal diameter with a heteroechoic lesion in upper pole. The abdominal computed tomography with contrast showed one zone of heterogeneous density in the spleen besides hemorrhagic liquid in peritoneal cavity, which confirmed the splenic rupture.

Splenectomy and a liver biopsy were performed. The patient's postoperative course was unremarkable. Pathologic examination of the spleen showed the splenic parenchyma densely infiltrated by lymphoid blasts; the normal architecture of the spleen had completely disappeared. Spleen blasts were positive for CD19 and CD20 monoclonals antibodies; liver biopsy showed leukemic infiltration of portal areas, with the blasts sharing the same immunophenotype.

A myelogram was performed concomitantly and acute lymphoblastic leukemia was diagnosed, L2 type, according to the FAB classification. The immunophenotype showed that bone marrow blasts were positive for CD19, CD20, CD22, CD10, CD34 and HLA-DR, and negative for CD3, CD4, CD5, CD7, CD13, CD14, CD15 and CD33. Bcr/abl translocation was not found. No central nervous system involvement was found. The patient has completed induction of a polychemotherapy regimen, BFM protocol,[8] with initially good response, and he has achieved complete remission after the induction therapy.

Giagounidis *et al.*,[1] have found that, apart from the three previously described pathogenic factors leading to rupture (splenic infiltration, splenic infarcts and coagulation disorders), male sex, adulthood, severe splenomegaly, and cytoreductive chemotherapy may be of relevance to the development of splenic rupture. The major pathogenic factor in this case seems to be splenic infiltration, since splenic infarcts and coagulation disorders were not found.

Altés *et al*.[2] tried to identify a possible association between pathologic spleen rupture with a particular genetic or phenotypic pattern. The findings encountered in their patient did not have any relevant diagnostic features. We have not either found any special immunophenotypic findings that could explain this particular form of presentation either.

In view of the results of Giagounidis series, splenectomy is advocated as the safest therapy. Nevertheless, Guth *et al*.[9] have recently proposed that a subset of hemodynamically stable patients can be successfully managed nonoperatively.

We present a further case of pathologic rupture of the spleen that warrants the need for differential diagnosis of hematological malignancies,[10] even though patient previous history do not suggest a hematological disorder.

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