shrinking of the pulmonary opacities, a complete radiological detersion having been achieved in September 1996 (Figure 2). At this time, just one year after starting it, cyclophosphamide was stopped. At the last control, in April 1997, the patient continued to be well, having normal routine laboratory analyses and negative chest radiography.

In the present case, both the lungs were affected by multiple neoformations, two of which were about 8 cm in diameter; to our knowledge, pulmonary masses of similar size have never been reported so far in a case of low-grade MALT-lymphoma, which is most often restricted to only one lung.[5,6]

It has been suggested that autoimmune diseases may play a pathogenetic role in the development of MALT-lymphoma.[2,7] Myasthenia gravis is also an autoimmune disease, characterized by an autoaggressive process directed against the acetylcholine receptors; however, an association between MALT-lymphomas and myasthenia gravis has never been reported so far. Whether a long-lasting myasthenia gravis may be considered an autoimmune disorder predisposing *per se* to the development of MALT-lymphoma remains an open question. However, a merely casual occurrence of two independent diseases cannot be excluded at present.

Optimal management of MALT-lymphoma is not well standardized yet. In spite of the extent of lung involvement, cyclophosphamide induced, in our patient, a progressive decrease of the pulmonary opacities, up to a complete radiological detersion one year thereafter. Clinical and radiological remission have now lasted for about seven months since the cessation of treatment.

Key words

MALT-lymphoma, myasthenia gravis, autoimmune disorders

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Mantle cell lymphoma with amyotrophic lateral sclerosis (motor neuron disease)

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We describe a previously unreported case of mantle cell lymphoma (MCL) associated to amyotrophic lateral sclerosis (ALS) in a 63-year-old woman with a 1-year history of weakness of arm and leg muscles. The both molecular-genetic and flow cytometry analysis of lymphocytes of peripheral blood (PB) demonstrated leukemic phase of MCL.

A 63-year-old woman was admitted to our hospital for one-year progressive weakness in her extremities. Tendon reflexes were overactive in the legs with widespread reflexing area, bilateral Babinski and both ankle and patellar clonus. There was no sensory defect. Brain magnetic resonance imaging showed cortico-subcortical atrophy and multiple small lesions of white matter. Electromyographic studies revealed severe multisegmentary denervation with preserved peripheral nerve conduction. A lumbar puncture showed cerebrospinal fluid protein content and biochemical profile were normal, Gram's stain and cultures were negatives. PB examination showed a white cell count of 19.5×10^9/L with an absolute lymphocyte count of 11×10^9/L. Hemoglobin level and platelet count were normal. The lymphocytes were medium to large in size, with scanty to moderate cytoplasm, and indented nuclei. The chromatin was moderately coarse with nucleoli in some cells. Serum immunoglobulin (IG) were normal, without monoclonal component. The immunophenotypic features of lymphoid cells were: $CD19^+$, $CD5^+$, $CD20^+$, kappa $(\kappa)^+$, lambda $(\lambda)^-$, $CD23^-$, $CD10^-$ and $FMC7^-$, with clonal expression of κ light chain on 64% of mononuclear cells. The bone marrow (BM) aspirate showed 37% mature-appearing lymphocytes. BM biopsy revealed reactive changes. Immunohistochemical study of BM specimen showed expression of both κ and λ chains of immunoglobulins. Cytogenetic analysis of BM showed no chromosomal abnormalities.

The body CT did not show hepatosplenomegaly, adenopathies or extranodal involvement. The endo-

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]

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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,