Case report

Idiopathic myelofibrosis with prominent postsplenectomy erythroblastosis terminating in acute myeloid transformation

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Abstract—Idiopathic myelofibrosis is a myeloproliferative disease with poor prognosis and without sufficient therapy. Acute leukemic transformation occurs in 15% of patients. The authors report the case of a 63 year old myelofibrotic patient treated with splenectomy. During the clinical course they observed unusually prominent and persistent erythroblastosis in the peripheral blood. After a two years long, relatively stable period the disease terminated in acute myeloid leukemia.

Key words: Acute leukemic transformation; idiopathic myelofibrosis; postsplenectomy erythroblastosis.

INTRODUCTION

Idiopathic myelofibrosis (IMF) is a clonal stem cell disorder characterized by chronic myeloproliferation with pleomorphic megakaryocyte hyperplasia, reactive bone marrow fibrosis, extramedullary myeloid metaplasia and peripheral leucoerythroblastosis with teardrop poikilocytes. Clinical features including splenomegaly, progressive anemia and constitutional symptoms. Treatment is largely palliative and median survival is 3–5 years [1]. Acute leukemic transformation occurs in about 15% of cases [2] but there are data on higher transformation rates with mixed leukemias according to stem cell origin [3]. Splenectomy may be necessary because of subjective abdominal complaints or obvious hypersplenism [4]. Morphological changes in peripheral blood after splenectomy are well known but prominent and persistent erythroblastosis is unusual [5]. We report on a myelofibrotic patient

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with long-lasting extreme erythroblastosis developed after splenectomy and a final myeloblastic transformation.

CASE REPORT

A 63 year old man was admitted to the hospital at the beginning of 1998 with abdominal pain, splenomegaly, icterus, and weight loss (10 kg per 3 months). His history comprised investigations and treatments between 1991 and 1994 because of pyelonephritis, duodenal ulcer and cholelithiasis. At that time the abdominal sonography showed a moderately enlarged spleen but there were no signs of alterations in hematological parameters.

Recent physical examination revealed splenomegaly (the spleen was palpable 10 cm below the costal margin). The laboratory tests showed signs of hemolytic anemia (hemoglobin: 105 g/l, reticulocyte: 40%, LDH: 538 U/l, bilirubin: 67.4 μmol/l, direct bilirubin: 10 μmol/l, the Coombs test was negative), leucopenia (2800/μl) and leucoerythroblastic smear with dacryocytes. The decalcified and paraffin-embedded iliac crest biopsy processed according to Schaefer [6] showed IMF: mature myeloid and pleomorphic megakaryocyte proliferation orientated by diffuse reticulin network, intrasinusoidal hemopoiesis, increased angiogenesis and less prominent erythropoiesis. Osteogenesis and blastic foci were not seen (Fig. 1). Otherwise, apart from traditional histological methods, we used several

Figure 1. Bone core biopsy at the time of diagnosis. ‘Streaming’ megakaryocytes characteristic of reticulin fibrosis (not shown) in idiopathic myelofibrosis. (Ulex europaeus lectin histochemistry, 300×).