**Case Report**

**Splenic Rupture in Acute Megakaryoblastic Leukemia**

**Pakming Wong, Katsuhiko Takabayashi, Yuri Sugiura, Takayoshi Asai, Kuniaki Itoh, Sho Yoshida and Hiroshi Horie**

A 43-year-old man with pancytopenia and circulating blast cells was found to have acute megakaryoblastic leukemia. Bone marrow biopsy disclosed reticulin fibrosis with infiltration of blast cells. Cytogenetic study showed 48,XY,+15,+18. The megakaryocytic origin of blast cells was confirmed by platelet peroxidase reaction at ultrastructural level. The spleen which had not been felt on admission increased in size rapidly and ruptured abruptly. This appears to be the first reported case of a splenic rupture associated with acute megakaryoblastic leukemia.

**Key Words:** Acute megakaryoblastic leukemia, Splenic rupture, Abdominal pain.

Acute megakaryoblastic leukemia (AMbL) is a relatively uncommon form of leukemia that is characterized by the megakaryocytic origin of the circulating blast cells. Overt splenomegaly was absent in most patients with AMbL, and spontaneous rupture of the spleen in this type of leukemia has not been reported. We describe a case of AMbL associated with splenic rupture.

**REPORT OF A CASE**

A 43-year-old man, a barber with a 6 week history of high fever and lumbago was found to have pancytopenia at a local hospital on April 15, 1983. Past history was unremarkable except for hyperthyroidism which was cured 5 years ago. There was no family history of malignancy. Physical examination on admission showed a well developed and wellnourished afebrile man. No lymphadenopathy or hepatosplenomegaly were noted. There were a few bruises and petechiae. His physical examination was otherwise unremarkable. Results of his hematological investigation were as follows: RBC 3,300,000 per mm$^3$, Hb 9.4 g/dl, platelet count 20,000 per mm$^3$, WBC 1,600 per mm$^3$ with 46% blasts, 8% promyelocytes, 2% myelocytes, 24% lymphocytes. Giant platelets were observed, but the erythrocyte morphology was normal. The blasts were moderate to large in size with a high nucleocytoplasmic ratio, a rich purple, moderately fine chromatin, a basophilic cytoplasm and for some cells, blebs on the plasma membrane. The blasts were positive for acid phosphatase but not for peroxidase, Periodic Acid-Schiff, naphthal AS-D chloroacetate esterase and alpha naphthyl butyrate esterase. The terminal deoxynucleotidyl transferase of the blasts was also negative. The blasts reacted antifactor VIII antibody. Electron microscopy showed undifferentiated blasts. Using ultrastructural cytochemistry, platelet peroxidase activity was demonstrated along the nuclear envelope and in strands of endoplasmic reticulum (Fig. 1). No myeloperoxidase activity was demonstrated in any blast cells. Bone marrow aspiration was attempted at multiple sites, but no particles were obtained. The trephine biopsy showed a hypocellular bone marrow, with prominent reticulin fibrosis. There was a marked infiltration of undifferentiated mononuclear cells in some areas of the marrow (Fig. 2). Cytogenetic studies of the blast cells showed a karyotype of 48,XY,+5,+18. Based on the above findings, a diagnosis of acute megakaryoblastic leukemia was made. The patient was treated with prednisolone (30 mg) daily. The clinical condition deteriorated rapidly with splenic enlargement, increased bone pain and high fever. On the 8th hospital day, the patient suddenly vomited and complained of increased abdominal pain. He became diaphoretic, hypotensive and had tachycardia. Over the next 12 hours, his hemoglobin...
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level fell to 4.8 g/dl, WBC increased to 21,400 per mm$^3$ with 58% blasts and the platelet count was 20,000 per mm$^3$. He had increased rebound tenderness and decreased bowel sounds. Ultrasonogram showed ascites and an enlarged spleen with low density areas. An intraabdominal hemorrhage was confirmed by paracentesis. A complication of splenic rupture due to rapid proliferation of the blast cells was considered. Blood transfusion was continued to maintain his blood pressure. In view of the patient's poor condition, vincristine (2 mg) weekly and prednisolone (30 mg) daily were given. On the 13th hospital day, WBC count decreased to 1,300 per mm$^3$, the abdominal pain had abated and his general condition was improved, which maintained for 3 months. At the beginning of Sep-

tember 1983, cytosine arabinoside (30 mg) was given for five days because of the increased bone pain. Pain was absent, but the general condition deteriorated with progressive pancytopenia. He died of pneumonia on the 130th hospital day. Au-

Fig. 1. Positive platelet peroxidase reaction in the nuclear membrane and endoplasmic reticulum of a circulating blast. (x6300)

Fig. 2. A Trephine biopsy section of bone marrow showing reticulin fibrosis with focal infiltration of undifferentiated mononuclear cells. Hematoxylin and eosin. (x100)

Fig. 3. The spleen showing numerous lacerations and infarcts.

Fig. 4. A histologic specimen of the spleen showing leukemic cell infiltration. Hematoxylin and eosin. (x200)
tomy revealed 1,140 gm spleen with numerous lacerations and infarcts (Fig. 3). There was leukemic involvement of the spleen (Fig. 4), liver, bilateral lungs and bone marrow.

**DISCUSSION**

The clinical and laboratory features of our case coincide with those of previously reported cases with megakaryoblastic leukemia: absence of overt hepatosplenomegaly or lymphadenopathy; pancytopenia with a few circulating blasts, normal erythrocyte morphology and giant platelets; diffuse reticulin myelofibrosis; multiple chromosome abnormalities; and poor response to chemotherapy. AMbL was first described by Von Boros and Korenyi in 1931. Confirmation of acute leukemia with megakaryocytic differentiation was difficult until Breton-Gorius and her colleagues identified the megakaryoblasts using ultrastructural cytochemical study of the platelet peroxidase reaction (PPO). Subsequently, several specific markers for megakaryocytes have been recognized, including factor VIII antigen, platelet membrane glycoproteins and acid phosphatase demonstrable by ultrastructural cytochemistry. The blast cells in our patient were positive for PPO and reacted antifactor VIII antibody. Since the methods for identification and confirmation of AMbL developed, reports on AMbL have been increasing during the past few years. Huang et al. reported 12 cases of acute leukemia with megakaryocytic differentiation, which represented from 11.8% of 85 cases with acute nonlymphocytic leukemia during a 12-month period of study. It seems that AMbL occurs more frequently than previously recognized, while the complications of AMbL are not well investigated. In our case, splenic rupture developed as an unusual complication, which has not been described in previous reports of AMbL. Splenic rupture is rare in patients with acute leukemia. Only six examples of splenic rupture were identified in 840 analyzed cases. The mechanism of splenic rupture in patients with acute leukemia remains uncertain. Leukemic infiltration and multiple infarcts found in our patient could have contributed to splenic rupture.

Severe abdominal pain along with hypotension, tachycardia, tachypnea and fever are the characteristic symptoms of splenic rupture. However, this pain was frequently confused clinically with that of acute leukemia and its other complications. We report this case as an unusual cause of an acute abdominal pain in AMbL. This appears to be the first case of a splenic rupture associated with AMbL.

**REFERENCES**


