Spontaneous remission in acute lymphoblastic leukemia: a case report and review of the literature

Abstract
Spontaneous remission (SR) has been reported in different hematological malignancies. Many cases of SR have been reported in acute myeloid leukemia (AML), few for childhood acute lymphoblastic leukemia (ALL) and rarely for adult ALL. An association of SR in acute leukemias with concomitant bacterial, viral or fungal infections has been noted. Spontaneous remission in adult precursor B lymphoblastic leukemia is rare. We herein, describe a patient with precursor B lymphoblastic leukemia seen at our institution that had spontaneous remission.

Keywords: spontaneous remission, precursor B lymphoblastic leukemia, hematological malignancies, coombs test, dyschezia

Introduction
Spontaneous remission or regression of cancer is defined as partial or complete disappearance of malignant disease temporarily or permanently in the absence of any specific treatment. The condition is named spontaneous regression for solid tumors and spontaneous remission (SR) for leukemia. Spontaneous remission has been reported in different hematological malignancies. It has been observed in adult T-cell leukemia/lymphoma, chronic lymphocytic leukemia and myelodysplastic syndrome. Many cases of SR have been also reported in acute myeloid leukemia (AML), few for childhood acute lymphoblastic leukemia (ALL) and rarely for adult ALL. An association of SR in acute leukemias with concomitant bacterial, viral or fungal infections has been noted. The cytokine release after infections has been considered to cause regression via immune modulation mechanisms. We herein, describe a patient with ALL seen at our institution that went temporarily for spontaneous remission.

Case presentation
A 19 years old female presented with fatigue and weight loss followed by fever, dizziness and vaginal bleeding started in October, 2014. The patient was admitted to the Hematology Unit, Oncology Center, Mansoura University in 21th, October, 2014. Laboratory tests showed anemia (Hb 5.37g/dL), leukopenia (WBC 1.75x10^9/L), severe neutropenia (0.048x10^9/L) and thrombocytopenia with platelet count 65.5x10^9/L. The differential leukocytic count showed 95.9% lymphocytes, 0.7% monocytes and 0.6% eosinophils with no blast cells detected in a blood film. The total bilirubin was 4mg/dl (direct 1.6mg/dl). Viral screen for HCV, HBV and HIV was negative and Coombs test was negative. The pelvi-abdominal ultrasound showed mild hepatosplenomegaly. Her bone marrow aspiration (BMA) was hypercellular with infiltration by 90% blast cells. The blast cells were heterogeneous in size with agranular rim of cytoplasm. The nucleus has fine chromatin with one to two nucleoli. No cytochemistry was performed but cMPO was negative by flow cytometry.

The diagnosis of precursor B lymphoblastic leukemia was confirmed by immunophenotyping on BMA which showed predominant B cell population (about 80% of all the cells analyzed). These B cells have small nuclear size (based on forward-scatter signal) with expression of CD10, CD19, CD38, HLA-DR and TdT. They are partially positive for CD20 and negative for CD2, CD3, CD4, CD8, CD5, CD7, CD56, CD34, CD13, CD117 and CD33. The blast cells were gated using CD45vs SS histogram on dim CD45 and low side scatter area. MPO was negative as well as monocytic markers like CD14, 64, 11c, 11b and CD4. BM chromosomal study showed no cytogenetic evidence of clonal abnormality with negative PCR for BCR-ABL transcripts (p190). The patient was planned for chemotherapy and started supportive measures. Two days after admission she complained of cough and dyspnea with crepitation and diminished air entry on the right side of the chest. Post-contrast CT chest revealed right sided consolidation with effusion. She complained of pain at the anal area and examination showed perianal wound and received local measures after consultation of surgery. Chemotherapy was postponed due to infection and persistent fever and the patient continued supportive treatment with hydration, antibiotics and transfusion of packed RBCs and platelet.

One week later, complete blood count showed more deterioration (WBCs: 0.9x10^9/L, neutrophils 0.046x10^9/L and platelet count 17x10^9/L) with persistence of fever. The antibiotics were modified according to the results of sputum culture and sensitivity. The lesion at the perianal area progressed with sever anal pain and dyschezia. Surgical consultation revealed perianal abscess eroding the anal canal with possible necrotizing fasciitis. One week later, left hemicolectomy with colostomy was done for necrotizing fasciitis. Her general condition started to improve with no fever with complete blood count showing recovery (WBC 2.7x10^9/L, neutrophil 1.7x10^9/L, platelets 190x10^9/L and Hb 11.7gm/dl). However, she was still not fit for chemotherapy from the surgical point of view. The patient was discharged with follow up with general surgery for dressing and supportive care. Two weeks later, her CBC showed complete recovery with WBCs 4.2x10^9, neutrophils 2.3x10^9, platelets 333x10^9 and Hb 12.1gm/dl. The blood film showed no evidence of circulating blast...
cells. The bone marrow was low normocellular with blast cells 2%. No records for BCR-ABL transcripts during remission. She was kept under monitoring and follows up only. One month later the bone marrow was normocellular with 2% blast cells.

Two months later, her clinical examination showed bilateral cervical lymph nodes and right inguinal lymph nodes of variable sizes (1-3cm) with leukocytosis and thrombocytopenia (WBCs 26x10^9/L, neutrophil 2.3x10^9/L, platelet: 61x10^9/L and Hb 12.2gm/dl). Peripheral blood smear showed blast cells and the immune-phenotypic was consistent with precursor B lymphoblastic leukemia. The patient lost follow up for 2weeks. In 19 March 2015, she had been admitted with fever, cough and vomiting with high serum creatinine (2.3mg/dl) and very high serum uric acid level (30mg/dl). The diagnosis of tumor lysis was established and she started hydration, uricosurics and antibiotics. Post-contrast CT chest revealed pulmonary nodules mostly infiltrations and bilateral consolidations mostly pneumonic.

After stabilization of her condition and improvement of her chemistry labs she received a prephase of chemotherapy consisted of vincristine 2mg day1 and 8mg dexamethasone/12hrs for 5days. She developed necrotizing fasciitis of the elbow region with severe pain and swelling of the elbow for which debridement occurred. However, she did not respond to the treatment with development of neutropenic fever and deterioration of her CBC (WBCs 0.62x10^9/L, neutrophil 0.48x10^9/L, platelets 7.9x10^9/L). She received antibiotics and antifungal according to the protocol of the hospital for neutropenic fever with transfusion support. Unfortunately, she developed septicemia and shock and transferred to ICU where she had been expired.

Discussion

Spontaneous remission in acute leukemia has been reported since 1878. Most of these cases were in the previous century. Nowadays, the effective chemotherapeutic agents have been widely available and are given immediately after establishing the diagnosis. Therefore, only a small group of patients as those with advanced age, poor performance status or presence of severe co morbidities was disqualified from small group of patients as those with advanced age, poor performance status or presence of severe co morbidities was disqualified from high risk of progression to acute myeloid leukemia (AML) M5b. Surgical biopsy and pneumonia was reported in a case of adult T-cell leukemia/lymphoma (ATL).2 Similarly, spontaneous remission in our case may be attributed to severe infections. A profound increase in the levels of cytokines such as tumor necrosis factor a (TNF-g), interleukin 2 (IL-2) and interferon-g (IFN-g) together with increased T-cell leukemia/lymphoma (ATL).

Discussion

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Conclusion

Spontaneous remission of ALL is rare especially in adults. Close follow-up is necessary as relapses will occur and spontaneous remissions are usually of short duration. Its mechanism is yet to be determined.

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None.

Conflict of interest

The author declares no conflict of interest.

References