
Massive Intraabdominal Relapse in a Patient with Acute Lymphocytic Leukemia

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ABSTRACT

Abdominal involvement in an isolated extramedullary relapse region in patients with acute lymphocytic leukemia (ALL) is a rare event, especially in patients in hematological remission. The literature related to this subject is very limited. In this study, a 23-year-old male patient, is presented who had been in remission for 4 years, was admitted because of an enlarged testis, and was found to have an abdominal bulky mass during physical examination. An early diagnosis of abdominal involvement is extremely important and must be kept in mind for ALL patients who are admitted for testicular relapse.

Key Words: Acute lymphocytic leukemia, Abdominal relapse, Testicular relapse.

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Extramedullary relapses in ALL patients generally occur in the central nervous system (CNS) and testis. Involvements in other sites generally occur during systemic relapses or multiple relapses in the late stages of ALL. Intraabdominal relapses in patients with ALL who are in hematological remission, rarely occur. In this paper we present the case of a patient with ALL who was in hematologic remission for 4 years, and had a abdominal and testicular relapse.

CASE REPORT

A 23-year-old male patient, complaining of an enlarged bilateral testis, was admitted to our hospital in October 1998. The patient had previously been treated in September 1994 for a diagnosis of CALLA (+) FAB-ALL1. Induction chemotherapy with vincristine, prednisone, daunorubicin, and L-asparaginase and consolidation with high-dose cytosine arabinoside had been given to the patient. A CNS prophylaxis had been given because the patient went into remission and later, maintenance chemotherapy was been given for 2.5 years. The patient had had no complaints since the

cessation of the maintenance chemotherapy 1 year previously.

Upon physical examination an irregular solid mass about 10x10 cm² was noted at the superior left quadrant of the abdomen. In addition, a painless mass was noted in the bilateral testis by palpation.

The laboratory data were as follows: hemoglobin: 15.9 gr/dL, hematocrit: 44.7%, WBC: 10.000/mm³ (%75 neutrophil), platelets: 296.000/mm³, erythrocyte sedimentation rate: 11 mm/h, LDH: 602 IU/L. A peripheral blood smear was normal and other biochemical tests and the urine analysis were within normal limits. The results of the bone marrow aspiration analysis showed that the patient was still in hematological remission. CEA, CA 19-9, CA 15-3, AFP, Beta-HCG, free PSA were normal. By testis ultrasonography (USG), a heterogeneous hypoechoic region (about 37x28 mm²) with an irregular shape in the right testis and a heterogeneous hypoechoic region covering all of the left testis (about 74x48 mm²) were detected. By computerized tomography (CT), a soft tissue mass, which covered the paraaortic aortacaval prevertebral region, extended into inferior about 14 cm, sized 11x19 cm² at the widest, was detected. This mass was interpreted as consistent with lymphadenopathy (LAP). The left ureter was inside the mass and seemed to be pressed. The left pelvis was dilated. A bilateral testicular biopsy showed bilateral ALL infiltration in the testicular tissue. A needle aspiration biopsy of the abdominal mass, performed during the CT, showed Class V lymphoblastic infiltration. No blast was detected in the cerebrospinal fluid (CSF).

Radiotherapy was used in treatment for the mass in the testis. The dose of radiation was 2400 cGy. At the same time, remission induction therapy with vincristine 2 mg/day IV on days 1, 8, 15 and 22, daunorubicin 45 mg/m²/day IV on days 1, 2, 3, cyclophosphamide 1000 mg/m² IV one dose in first day, L-asparaginase 6000 IU/m²/day IV starting in 5th day, 2 times a week for a total of 6 doses, and prednisone 60 mg/m²/ on days 1 to 29 PO were given. Also, during this therapy, 2 times a week and total of 6 doses of intrathecal methot-

rexate were given.

After treatment, an analysis of the bone marrow aspiration and testicular and abdominal USG were repeated. Hemotological remission was still continuing. In the testis and the abdominal USG, there was no significant shrinkage in the dimension of the mass. Abdominal radiotherapy was planned, but could not be performed, due to the rapid deterioration in the general condition of the patient. Unconsciousness and muscle weakness developed in the patient and millimetric lesions, consistent with cranial relapse, were observed by MRI examination. The patient died in a couple of days.

DISCUSSION

After the introduction of effective systemic chemotherapy for the treatment of ALL, relapses in the sanctuary site occurred due to the prolongation of survival. Testis and central nervous system leukemia are the most common forms of extramedullary relapse^[1]. Other than these sites, isolated visceral relapses are not common. Leukemic visceral infiltration usually occurs in association with a massive bone marrow involvement^[2].

Individual case reports and autopsy data indicate that in acute leukemia cases many viscera may be infiltrated with leukemic cells as asymptomatic^[1,3,4]. However, in patients with ALL in hematological remission, massive intraabdominal relapse is exceptional^[1]. In case reports as well as in our case, the importance of an early diagnosis and treatment for abdominal relapse is emphasized in order to prolong survival^[5,6].

In the study of Edward Baum and Associates, 6 patients are evaluated by laparotomy. In three of these six patients, occult abdominal nodal involvement was observed. Chemotherapy and testicular treatment were performed on all three of these patients. In addition to this therapy, total abdominal RT was given to only one of these patients. Among three of the patients, only the one who was given RT survived^[7]. In our case bulky disease was present in the abdomen. Systemic chemotherapy and bilateral testicular RT were given. However, the results of this therapy was not encouraging. Abdominal radiotherapy was plan-

ned, but could not be performed due to the rapid deterioration in the general condition of the patient. After developing intracranial involvement, the patient died. The prognosis deteriorated more in this case due to bulky disease. Hence, the importance of early diagnosis was clear. It seems that total or nodal abdominal RT, in association with systemic chemotherapy, is the most suitable therapy, especially in early diagnosed abdominal relapse.

If testicular relapse occurs early during the course of therapy, it is a bad prognostic sign and is usually followed by marrow relapse. But testicular disease occurring late, especially after elective cessation of therapy, is compatible with long-term disease-free survival following appropriate therapy^[9]. In our case, relapse occurred in a testis one year after cessation of maintenance therapy. However, abdominal involvement, in addition to testicular disease, caused a change in the prognosis. In this regard, the possibility of relapse must be kept in mind in patients with ALL who have testicular relapse. The studies on the roles of tomography and USG in occult abdominal involvement are unsatisfactory^[8]. In addition the role of lymphangiography in early diagnosis is not yet known. We believe that patients with acute leukemia should be followed by noninvasive procedures such as abdominal USG, especially during remission. Therefore, comprehensive studies are needed on this subject.

REFERENCES

1. Hustu HO, Aur RJA. Extramedullary leukemia. Clin Haematol 1978;7(2):313-37.
2. Silverstein MN, Bayrd ED. Nonmeningial extramedullary relapse in leukemia. Arch Int Med 1969; 123:401-4.
3. Cornes JS, Jones TG. Leukemic lesions of the gastrointestinal tract. J Clin Pathol 1962;15:305-13.
4. Mathe G, Schwarzenberg L. Extensive histologic and cytologic survey of patients with acute leukemia in complete remission. Br Med J 1966;20:640-2.
5. Uderzo C, Santamaria M. Abdominal mass as manifestation of isolated extramedullary relapse in a child with acute lymphoblastic leukemia. Haematologica 1987;72:545-7.
6. Kurtz JE, Andres E. Intraabdominal localization of acute leukemia. Nouv Rev Fr Hematol 1995; 37:273-6.
7. Baum E, Heyn R. Occult abdominal involvement with apparently isolated testicular relapse in children with acute lymphocytic leukemia. Am J Pediatr Hematol Oncol 1984;6:343-5.
8. Geoffroy A, Shirkhoda A. Abdominal and pelvic computed tomography in leukemic patients. J Comput Assist Tomogr 1984;8(5):857-60.
9. Russo A, Schiliro G. The enigma of testicular leukemia. Med Pediatr Oncol 1986;14:300-5.

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