Fatal intramedular tumor of the cervical spinal cord during remission of acute lymphoblastic leukemia

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Summary

An adult male with acute lymphoblastic leukemia including meningeal leukemia was in complete clinical remission for more than one year after initiation of chemotherapy including intrathecally methotrexate and irradiation towards the brain and spinal cord. Signs of central nervous system involvement developed with symptoms primarily from the lower limbs and the urinary bladder but the cerebrospinal fluid was consistently without lymphoblasts. By magnetic resonance imaging an intramedular tumor of the cervical spinal cord was demonstrated and malignant cells compatible with lymphoblasts were obtained from the tumor at laminectomy. Despite immediate irradiation towards the tumor the patient died within two weeks.

Introduction

Clinically significant spinal cord involvement is unusual in leukemia, although meningeal involvement is common in the acute leukemias. When present, tumors extrinsic to but compressing the cord are the most common finding and the thoracic cord is the most common site of involvement [1]. However, with the increasing ability of intensive chemotherapy protocols to induce and maintain hematological remission it can be expected that development of late complications inside the central nervous system will emerge. I here report the clinical course of an adult male with sustained first clinical remission of acute lymphoblastic leukemia, who developed an intramedular spinal tumor which proved to be fatal due to the unfavorable anatomic localization in the upper spinal cord.

Case report

A 17-year-old male developed fever, dyspnea and hemorrhagic diathesis in March 1986. Pericardial effusion was found on echocardiography. CT scans showed splenomegaly and mediastinal lymphadenopathy. His initial leukocyte count was $95 \times 10^9/\text{l}$, the hemoglobin was $5.5 \text{mmol/l}$, and the platelet count was $10 \times 10^9/\text{l}$. A bone marrow biopsy revealed acute lymphoblastic leukemia (FAB type L1) with 96% blasts with T-cell surface markers. Cytogenetic examination showed a normal karyotype. Magnetic resonance imaging (MRI) showed a prolonged T(1)-relaxation time of the bone marrow. Induction therapy with prednisone, vincristine, adriamycin and L-asparaginase resulted in complete hematological remission, which was further consolidated with repetitive courses of 6-mercaptopurin, methotrexate, cyclophosphamide, cytosine arabinoside, vincristine and prednisone. Lumbar puncture in May 1986 revealed meningeal acute lymphoblastic leukemia of the same type as previously found in the bone marrow. Treatment consisted of intrathecal methotrexate (six doses) and irradiation of the brain and spinal cord (totally...
Fig. 1. Magnetic resonance imaging of the spinal cord of a patient with acute lymphoblastic leukemia in complete hematologic remission. An intramedular tumor (arrows) is shown at the level of cervical spine C3-C4.

24 Gy), after which no leukemic blasts could be found in the cerebrospinal fluid.

The patient remained in complete remission for 15 months until 1st September 1987, where progressive weakness of the legs and urinary incontinence appeared. The cerebrospinal fluid was without lymphoblasts and a myelogram was normal. On neurologic examination the patient was lethargic and with moderate paresis of the lower limbs. The deep tendon reflexes were increased and plantar responses were bilaterally extensor. Symmetrically decreased sensory senses were found. Within a week rapid progression to complete paralysis of the lower limbs, partial paresis of the left upper limb, and paresis of the urinary bladder developed. The cerebrospinal fluid was consistently without blasts and the bone marrow normal. However, MRI of the CNS revealed a solid spinal tumor of the cervical cord (Fig. 1). Acute laminectomy of the cervical spine was performed, but radical resection of the tumor was not possible. Histopathological examination showed malignant cells not inconsistent with lymphoblasts, but severe necrosis hindered an exact cytological diagnosis. However, there was no evidence of other neoplastic cells, and there was no signs of intraspinal hemorrhage at operation. Abscess was ruled out, since no bacteria could be demonstrated upon microbiological examinations. Postoperative irradiation (totally 16 Gy) was given towards the intraspinal tumor, but although a temporary improvement was observed further expansion of the intramedullar leukemic tumor took place, paralysis of the upper and lower limbs developed and the patient died after two weeks due to general seizures. Permission for autopsy was not obtained.

Discussion

In acute lymphoblastic leukemia intraparenchymal leukemic masses of the central nervous system are uncommon, although the incidence may be increasing as more and more patients achieve com-