K. Krishnan · C.W. Ross · P.T. Adams · A. Pereira
M.S. Roth

Neural cell-adhesion molecule (CD 56)-positive, t(8; 21) acute myeloid leukemia (AML, M-2) and granulocytic sarcoma

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Abstract A 49-year-old man with t(8; 21) acute myeloid leukemia relapsed 8 months after successful induction chemotherapy with a paraspinal granulocytic sarcoma. There was no evidence of leukemia in the bone marrow at relapse. At initial presentation, the blasts co-expressed CD 15, CD 33, CD 34, CD 45, CD 19, and CD 56 (a neural cell-adhesion molecule). Expression of certain cell-adhesion molecules on leukemic blasts may determine a tendency to develop extramedullary relapse. The co-expression of CD 56 may have a role in the predisposition of t(8; 21) AML to develop GS.

Key words t (8; 21) AML · Granulocytic sarcoma CD56 · Adhesion molecule

Introduction

Granulocytic sarcomas (GS) are extramedullary leukemic deposits seen in acute myelogenous leukemia (AML), myeloproliferative disorders, and myelodysplastic syndromes. A patient with t(8; 21) AML relapsed with GS in the paraspinal soft tissues following successful induction chemotherapy. We postulate that the increased incidence of GS in t(8; 21) AML may be related to the co-expression in leukemic blasts of CD 56, a neural cell-adhesion molecule (NCAM). Adhesion molecules represent homing receptors with an important role in targeting of neoplasia to particular types of tissue and determining patterns of metastasis [8].

Case report

A 49-year-old man presented in October 1992 with submandibular lymphadenopathy and pancytopenia. Laboratory tests revealed a white blood cell count of 3.3 x 10^9/l, hemoglobin of 10.5 g/dl, platelet count of 23 x 10^9/l, and an elevated serum lactate dehydrogenase of 560 IU/l. A bone marrow biopsy showed replacement by myeloblasts and the presence of Auer rods in some blasts. The leukemic blasts were strongly positive for myeloperoxidase and negative for non-specific esterase. Flow cytometry of the bone marrow blasts (Table 1) showed co-expression of CD 15, CD 33, CD 34, CD 45 and CD 56, weak expression of CD 7, and aberrant expression of CD 19 (B-lineage-associated antigen). Cytogenetic analysis revealed two populations of cells. One population (3/20 cells) contained a terminal deletion of the X chromosome and a translocation between the long arms of chromosomes 8 and 21. The second population (17/20 cells) had these abnormalities as well as a translocation between the short arm of chromosome 7 and the short arm of a second derivative chromosome 8 (Table 1).

The patient received induction chemotherapy with cytosine arabinoside (100 mg/m^2 on days 1–7) and daunorubicin (45 mg/...
m² on days 5–7) in November 1992. Bone marrow examination on day 30 following induction confirmed complete remission. He then developed non-neutropenic fever and multiple pyogenic lesions in the liver requiring prolonged administration of parenteral antibiotics. Cultures from a liver biopsy grew a mixture of organisms consisting of Enterobacter and coagulase-negative staphylococci. Subsequently, he developed Pseudomonas aeruginosa infection at an indwelling catheter site. It was elected to delay consolidation therapy because of these infections. A repeat bone marrow examination in March 1993 was normal.

Computerized tomographic (CT) scans of the chest and abdomen performed in May 1993 revealed a mass in the posterior mediastinum with extensive left paravertebral abnormal soft tissue from approximately the fifth to the tenth thoracic vertebra. Lumbar puncture and cerebrospinal fluid analysis were negative for malignant cells. He underwent thoracotomy and multiple biopsies of this mass. The histopathology (Fig. 1) and immunohistochemistry were diagnostic of a granulocytic sarcoma. Immunohistochemical stains on the neoplastic cells were positive for common leukocyte antigen and CD 43, with focal muramidase (lysozyme) positivity. The neoplastic cells were negative for cytokeratin (CAM 5.2), CD3, CD 20 (L 26), CD 15 (LeuM 1), and CD 45RO (UCHL1). In addition, a Leder stain was performed on formalin fixed tissue, which showed occasional chloroacetate esterase-positive cells. Wright’s stain on an imprint of the biopsy revealed occasional blasts with Auer rods (Fig. 2). Bone marrow examination at the time of extramedullary relapse was free of leukemia. In August 1993, the patient received intrathecal methotrexate prophylactically and underwent syngeneic bone marrow transplantation. His course following transplantation was complicated by multisystem failure, fluid retention, and eventual sepsis and death in September 1993.