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S Mirvis, M Stewart and K C Rao


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Myelographic Demonstration of "Nodular Radiculopathy" in Acute Myelogenous Leukemia

Stuart Mirvis,1 Michael Stewart,2 and Krishna C. V. G. Rao1

Documented central nervous system (CNS) involvement in acute myelogenous leukemia (AML), unlike acute lymphocytic leukemia, is relatively uncommon [1, 2]. CNS involvement in AML is reported to occur in 6%-17% of cases [1-4]. It is expected that improvements in therapy with increased frequency of remission and prolonged survival will lead to further increases in clinical CNS involvement in AML [1, 2].

A patient with documented acute myelomonocytic leukemia complained of symmetric, bilateral radicular symptoms while considered to be in clinical remission. A Pantopaque myelogram revealed bilaterally symmetric nodular masses involving the C8 nerve-root sleeves. Follow-up study after local radiation therapy demonstrated complete resolution of these findings by both computed tomography (CT) and repeat myelography.

Case Report

A 41-year-old man was diagnosed with acute myelomonocytic leukemia in October 1979. He underwent remission induction therapy 1 month later with doxorubicin (Adriamycin) and cytosine arabinoside (Ara C-Cytosom). The cerebrospinal fluid (CSF) was normal, and cytologic analysis revealed no evidence of CNS leukemia. No CNS prophylactic treatment was given. Complete bone marrow remission was documented 27 days after therapy was begun.

In June 1981 he presented with neurologic symptoms involving the lumbar and sacral regions. Evaluation disclosed leukemic meningitis with 1600 cells/mm² in the CSF (all myeloblasts on cytologic analysis; protein, 100 mg/dl; and glucose, 39 mg/dl). The patient received intrathecal cytosine arabinoside via lumbar cistern injection and later intrathecal methotrexate with external beam irradiation given to include the T12–S1 region. All symptoms were relieved, but blast forms persisted in the CSF. Subsequently, he developed cranial polyneuropathies involving primarily the bulbar region. These were believed to be from leukemic infiltration, and irradiation to the posterior fossa was undertaken with resolution of symptoms. The CSF was clear of all cells with normal chemistries after treatment with intrathecal methotrexate via Ommaya reservoir.

In December 1981, he developed cervical pain and proprioceptive dysfunction with mild signs of increased intracranial pressure believed to be from CNS involvement with leukemia. The CSF was unchanged with no evidence of malignant cells. He received dexamethasone and whole-brain irradiation. Again all symptoms resolved quickly and dramatically with irradiation. Systemic chemotherapy, however, was continued, and the patient was functional and well.

In August 1982, he developed bilateral shoulder pain extending posteriorly down the arm to the wrists. The pain was initially thought to be from local muscle strain but worsened over the next 2 weeks. Mild motor weakness developed in the right hand with reduced oppositional ability. Examination of the CSF was unrevealing. Clinical examination suggested a focus in the lower cervical spine region. Electromyography showed definite abnormality in the C7 or C8 nerve roots.

A Pantopaque myelogram showed symmetric, rounded, bilateral enlargement of the C6 nerve-root sleeves (fig. 1A). In light of the patient's history, it was elected not to explore the lesions surgically but to treat again with irradiation watching for a response to low dose. Indeed, treatment with 500 rad (5 Gy) in two fractions in the C6–T2 region produced dramatic relief of symptoms with elimination of pain and paresthesia and marked improvement in motor function. Repeat myelography and spinal CT 6 weeks later revealed resolution of previously noted root-sleeve defects (fig. 1B). The patient was still in bone marrow remission after more than 3 years.

Discussion

Clinically recognized involvement of the CNS in AML has been reported to occur in 6%-17% of cases [1-4]. Before 1970 clinically apparent CNS involvement was rarely reported, with only a single case in 200 consecutive patients [1]. The incidence has increased since the advent of more aggressive and successful chemotherapy regimens with substantially higher remission rates and improved survival [1, 2].

Autopsy studies have demonstrated that the incidence of CNS infiltrate in AML is greater than suspected clinically [1-4]. Wolk et al. [3] reported a 6.5% incidence of clinical CNS disease, while 19% of their patients had autopsy-proven CNS involvement. Similarly, a 26% incidence of "marked" CNS infiltrate was reported by Reske-Nielsen et al. (cited in [2]). In one autopsy study the incidence of infiltration of the dura was as high as 59% [4]. Clearly, clinical evaluation has tended to underestimate the presence of CNS disease in AML.

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1 Department of Diagnostic Radiology, University of Maryland Hospital, 22 S. Greene St., Baltimore, MD 21201. Address reprint requests to K. C. V. G. Rao.
2 University of Maryland Cancer Center, Baltimore, MD 21201.
to the formation of focal tumors in relation to neural structures, particularly the spinal cord, confined to granulocytic leukemia. They reported three instances of radicular syndromes with nerve-root invasion and two cases of cord compression based on neurologic and CSF findings without radiologic confirmation. These syndromes were not necessarily accompanied by leukemic meningitis. They suggested that focal tumors may have a special predilection for the CNS in both acute and chronic granulocytic leukemia.

Pippard et al. [4] described five (8%) of 64 patients with clinically evident CNS disease. Two of the five had myelomonocytic leukemias. Similarly, Ruggero et al. [2] reported five patients with CNS involvement in acute myeloid leukemia in a series of 55 patients. Again, two of six patients with myelomonocytic, but only three of 19 with typical myeloblastic, leukemia had evidence of CNS disease. Thus, this particular, rare variant of granulocytic leukemia may demonstrate a greater tendency to produce CNS disease than other subtypes.

Our patient also had the myelomonocytic variant of acute granulocytic leukemia. He presented with peculiar symptoms of bilateral radiculopathy after a prolonged period (8 months) of CNS and bone marrow remission. Despite normal CNS findings, myelography clearly demonstrated radicular involvement with nodular infiltration at the C8 nerve roots corresponding to the patient’s clinical symptoms and electromyographic abnormalities. Low-dose radiation therapy directed to this level produced complete and rapid resolution of symptoms with disappearance of the initial myelogram findings. Similarly, Pippard et al. [4] reported dramatic response of local leukemia CNS infiltrates in AML with radiotherapy or intrathecal cytotoxic drugs, provided it was not associated with uncontrolled granulocytic leukemia.

It is not clear why this patient developed spinal nerve-root infiltration despite previous extensive chemotherapy via Omaya reservoir and lumbar puncture with clearance of the CSF. Perhaps the spinal root sleeves present a physical barrier to the diffusion of chemotherapeutic drugs in the CSF and in some manner represent a "sanctuary within a sanctuary." In a patient with AML, especially the myelomonocytic variety, presenting with unusual complaints of a radicular nature, radiologic evaluation with myelography and metrizamide CT should be undertaken even with a normal CSF examination to exclude spinal root involvement.

With the availability of safer, water-soluble, nonionic myelographic contrast agents in combination with CT, the chance of detecting focal leukemic infiltrates, either as focal masses or infiltrating lesions involving the nerve roots, appears significantly better. Since prolonged remission and survival appear possible with newer treatment methods, it appears reasonable to pursue diagnostic studies in appropriately selected patients.

REFERENCES

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