

CASE #7

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Reference: two slides

The patient was an 8-year-old white male, well until age 4 when he was diagnosed as having acute lymphocytic leukemia. His initial remission was induced with vincristine, prednisone and central nervous system irradiation with 2400 rads, as well as intrathecal methotrexate. He was then maintained on 6-mercaptopurine, methotrexate and Cytosan with his remission lasting two years. Subsequent to that time, the patient had had numerous and increasingly frequent relapses, both of the bone marrow and the central nervous system. The patient was admitted for bone marrow transplantation from his HLA matched older brother. He was placed on the SCARI regimen and given total body irradiation with the marrow transplantation

Subsequently, he suffered numerous episodes of fever and chills but cultures remained negative. The hematologic parameters and general condition continuously improved, for the next 2 1/2 weeks. when he first became hallucinatory. Over the ensuing week, he had occasional brief episodes of lucid mental status, but generally his CNS function progressively decreased to the point of a flat line EEG on the day of his death, 9 days later.

Autopsy findings: There was bone marrow hypocellularity with marked myloid predominance, lymphoid depletion and severe thymic atrophy. Lungs showed pulmonary edema, focal atelectasis and focal hemorrhagic pneumonitis. Kidneys were edematous with focal acute tubular necrosis.

The central nervous system revealed periventricular and bilateral putaminal necrosis.

Microscopic slides: 1. H & E - Lateral and medial thalamus, putamen.
2. PAS - Pons.

Points for discussion:

1. Five (or more!) differential diagnoses should be considered in the evaluation of this case.
2. Which nosologic diagnostic entity is most appropriate?