Case 2006-2

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The patient was a 20-year-old male with a four year history of chronic myelogenous leukemia who was admitted for a third bone marrow transplant after relapse. He was admitted for conditioning with Busulfan, Fludarabine and Campath following which he received a 4/6 HLA matched related donor peripheral blood stem cell transplant. The transplant failed to engraft and he subsequently developed extreme neutropenia and recurrent fever unresponsive to various combinations of antibiotics. Fever was associated with headache and neck stiffness. Initial CSF analysis revealed no pleocytosis or significant alteration in CSF glucose and protein. CSF cultures failed to yield any organisms. His neck stiffness continued and later progressed to confusion followed by a state of “CNS depression”. CT scans of chest, abdomen and pelvis failed to reveal any significant abnormalities. CT and MRI of the brain showed findings suggestive of early communicating hydrocephalus. He continued to receive antibiotics. Due to failure to engraft and persistent mental status changes, DNR orders were written after consultation with family. He remained unresponsive to therapy and died after hospitalization for 5 weeks. Consent for an unrestricted autopsy was given. Significant findings included hemosiderosis secondary to chronic transfusion therapy, lymphoid depletion in the lymph nodes and spleen, cutaneous features of graft versus host disease and acute tubular necrosis with focal regenerative changes. The brain was swollen with prominent uncal grooves, bilaterally. A whitish discoloration was noted along the leptomeningeal vessels. The lateral ventricles contained a slimy material associated with slight greenish discoloration of the choroid plexus.

Material submitted: Slide: One H&E stained section of cerebral hemisphere or medulla and one unstained slide

Discussion:

What is the diagnosis or possible differential diagnosis?

What additional studies would you recommend to confirm diagnosis?