## Case 5

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Clinical Abstract:

The patient was a 14-year-old boy admitted to the hospital with a two-week history of nausea, abdominal pain, diarrhea, headaches, myalgias, and lethargy. Examination revealed tender cervical and axillary adenopathy, exudative pharyngitis and marked hepatosplenomegaly. A peripheral blood smear showed lymphocytosis with many atypical forms. Liver function tests were abnormal and a mono-spot test was positive. Epstein-Barr virus capsid antigen (VCA) titer was 1:320 with an IgM fraction of 1:80.

While in the hospital, the patient developed severe pancytopenia and a bone marrow examination showed no erythroid or granulocyte precursors, decreased number of megakaryocytes, and marked plasma cell infiltration. Acylovir, steroids and blood transfusions were given. Four weeks after admission, he developed lower G.I. tract hemorrhage and a few days later presented generalized seizures. On neurologic examination papilledema was noted but no focal signs could be demonstrated. CT scan showed mild hydrocephalus and a lumbar puncture yielded a fluid at an opening pressure of 300, with increased proteins, moderately low glucose level, and 50 atypical lymphocytes per µl. Immunocytochemistry revealed a polyclonal population. Subsequently, his hospital course was complicated by severe pulmonary edema and hypotension requiring permanent respiratory support and vasopressors. The patient continued having frequent episodes of generalized seizures, his mental status deteriorated markedly and he developed left hemiparesis. Terminally, he had severe renal insufficiency and anasarca. He expired five weeks after admission.

Necropsy findings included necrotizing nodules in multiple organs (lungs, liver, testes, gastrointestinal tract, and lymph nodes) and thymic atrophy. The brain was markedly swollen with a light green discoloration, innumerable petechial hemorrhages, and small foci of necrosis throughout the cortex, deep gray matter and hemispheric white matter. Similar changes were noted in the cerebellum, brainstem, and spinal cord.

Material submitted: One H&E stained section from the right hippocampus or right basal ganglia

Points for discussion:
1. Diagnosis
2. Pathogenesis