43rd Annual Diagnostic Slide Session 2002

Case #2002- 8

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Clinical History: The patient was a 48 year old male who was admitted to this hospital for agitation and confusion. Past history reveal that at age 41 he was found to have mediastinal lymphadenopathy but refused treatment until one year later when more symptoms occurred. A lymph node biopsy at that time revealed nodular sclerosing Hodgkins disease. He was treated with ABVD and achieved a complete remission as measured by Gallium scan after six or eight cycles. Mantle radiation was begun but the patient stopped treatment after 3 days because of chest tightness. At age 43 there was recurrent disease manifested by mediastinal lymphadenopathy but he did not receive treatment until two years later when periaortic nodes were positive on Gallium scan. He was placed on ifosfamide, Etoposide and carboplatin and underwent an autologous stem cell transplant the same year. The following summer, the patient developed pruritis and restaging revealed lymphadenopathy. A groin lymph node biopsy was consistent with recurrent Hodgkin's disease. He was put on procarbazine 100 mg and Leucoran a day to control the disease which was considered incurable with some progression of his disease. That same year he received 3750 cGY of radiation therapy to the inguinal and femoral areas. One year later the patient was supposed to receive more radiation therapy but it is not clear if he received it. Late in the fall he was placed on gemcitabine and Navelbine therapy but he felt it was too toxic so did not continue it. He was seen here with some weight loss, fevers and persistent sweats. He was placed on procarbazine 100 mg a day and Advil p.r.n. He also took prednisone in an unknown dose. A recent CT scan of the chest demonstrated no residual adenopathy of the mediastinum; however, there was significant enlargement of axillary nodes. He was doing relatively well on unknown doses of Prednisone until this admission and had refused other treatment.

Social History: Family history is unknown. The patient lived alone and was unmarried. The patient has been unemployed and disabled secondary to back injury for several years.

Medications: Prednisone of unknown dose.

Physical Exam: BP was 100/60 after sedation. Pulse 92, respirations 18, temperature 98.6, Pulse ox 100 % on room air. The patient was awake but lethargic due to sedation and oriented x1. Pupils were round, regular and equal and reactive to light and accomodation. Extraocular movements were intact. The rest of the HEENT was negative as was the rest of the physical examination

Neurological Examination: The patient was responsive only to pain due to sedation and reflexes were absent. Babinski signs were negative.

Laboratory Studies: Hemoglobin was 10.4 and platelets 96000. White cell counts were normal except for low lymphocytes and high monocytes. Head CT scan was negative for hemorrhage and head MRI with and without contrast was normal. EKG showed a normal sinus rhythm. Chest x-ray showed mediastinal lymphadenopathy but otherwise was clear. The spleen was slightly enlarged. Urine tox screen was negative. EEG showed mild diffuse slowing on sedation with no epileptiform activity. Electrolytes were normal except for intermittent increased BUN and Creatinine and other metabolic studies showed an intermittently increased Na but were otherwise negative. Bilirubin was normal but there was consistently high AST, Alkaline phosphatase and LDH. CSF examinations were done several times and were negative for bacteria, fungus, virus, and TB. CSF showed a negative cytology for tumor. VDRL was negative although protein was elevated at 68 mg/dl. One urine culture showed multiple colonies of Proteus mirablilis sensitive to fluoroquinolones. On Gram stain of sputum showed moderate numbers of Staph aureus and Proteus mirablilis.

The patient was hospitalized for several weeks before death. Multiple anaerobic and aerobic blood cultures were negative. He was not on any immunosuppressive drugs in the hospital. Multiple CSF exams, MRI's, metabolic studies were negative except as indicated. He continued to deteriorate to lethargy, coma and death.

General autopsy findings showed only recurrent Hodgkins disease in axillary, retroperitoneal, mediastinal and periaortic lymph nodes as well as in liver, spleen, kidney and bone marrow. The entire brain was affected by the process you see on your slides. Gross examination revealed nothing.

Materials: H&E slides of cerebral or cerebellar white and gray matter or brainstem

Question: 1. What is the diagnosis?
2. What is the pathogenesis?