CASE 6

Submitted by: Debra B. Novotny, M.D., Frederick G. Dalldorf, M.D., and Kinuko Suzuki, M.D.
Department of Pathology
University of North Carolina
Chapel Hill, North Carolina 27514

Clinical Abstract:

The patient, a 75 year old man, was in excellent health until three years antemortem when he began to note progressive bilateral multiple joint pain and swelling. He was initially treated with NSAID, but he subsequently required Prednisone therapy for approximately one year antemortem. His past medical history was notable for >100 pack years of cigarette smoking.

He presented to clinic 2 months antemortem with a 6 month history of progressive general malaise, anorexia, dyspnea on exertion and a 60 pound weight loss. He denied fevers, chills or night sweats. A chest x-ray revealed a poorly-defined nodular density in the lateral right upper lobe. The lung fields were otherwise clear. An EGD revealed candidal esophagitis attributed to chronic steroid therapy. He was treated with a 2 week course of oral Nystatin.

He was last admitted 6 weeks antemortem for further evaluation of weight loss. A repeat EGD showed resolution of the esophagitis. He was anergic to PPD and control skin test antigens. An abdominal CT scan revealed only pleural and pericardial effusions. An ECG showed first degree AV block and bundle branch blocks. Shortly after admission, he had an episode of high anterior chest pain of 30 minutes duration without radiation or diaphoresis. A repeat ECG showed second degree AV block necessitating placement of a pacemaker. A myocardial infarct was ruled out by serial cardiac isoenzymes. High resolution electrocardiography revealed possible intra-Hisian disease. An echocardiogram showed fused, thickened aortic valve cusps and a moderate to large pericardial effusion.

He continued to have recurrent episodes of atypical chest pain attributed to angina, pericarditis, or recurrent esophagitis. Serial chest x-rays showed persistent bilateral effusions and atelectasis. He subsequently had a fatal cardiorespiratory arrest.

The patient had no clinical signs or symptoms of neurologic disease throughout his course. The brain weighed 1430 gm. The cerebral dura and falx cerebri were thickened and had focal nodularity. There was diffuse, firm, homogeneous, yellow-white thickening of the leptomeninges along the medial surfaces of the cerebral cortex adjacent to the falx.

Material submitted: One H&E and one unstained slide from the left medial parietal lobe

Points for discussion: 1. Diagnosis

2. Associated systemic findings
3. Clinicopathologic correlations