CASE 9

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Clinical Abstract:

This patient was a 44 year old White male who was well until early 1977, when he was hospitalized for an upper respiratory infection and was told that he had "low blood count". In June, 1978, he developed dyspnea on exertion and easy fatigability. Bone marrow examination revealed pancytopenia. He was started on Folate. Repeat bone marrow studies also showed hypocellularity. His family history was remarkable in that he had a brother who died of aplastic anemia complicated with pneumonia at age 47. Our patient was seen in June, 1979, at the Mayo Clinic, where a diagnosis of familial aplastic anemia was made. He was started on halotestine 30 mg. daily and prednisone 20 mg. daily.

On admission to KUMC he had bilateral retinal hemorrhages and petechiae over the extremities and trunk. He also had many white patches of the oral mucosa. He had a hemoglobin of 7.4 gm., 1,000 white blood cells and 15,000 platelets. Chest x-ray showed diffuse fibrosis and bilateral patchy pneumonitis of both lungs. Suppressor T cell function studies were done but results were not obtainable. Upper G.I. studies were consistent with moniliasis of the esophagus (multiple superficial ulcers). Nystatin and amphotericin B therapy was begun. The patient developed macular, well-circumscribed, crusted lesions periorally, and had fever, chills and weakness. Four days later acute changes in mental status were noted, which included drowsiness and receptive aphasia. Later that day the patient had a grand mal seizure and afterwards was unresponsive, with flaccid paralysis and doll's eye sign. EEG showed diffuse slowing. Lumbar tap showed clear spinal fluid, glucose 114 mg%, protein 25 mg%, no cells, negative Gram stain and India ink preparation. The patient expired the next day.

Necropsy Findings:

Bone marrow was almost acellular. There were esophageal ulcers and necrotizing pneumonia with masses of aspergillus organisms. The brain was swollen and contained multiple areas of greyish softening, ranging in size from 1 to 8 mm., in cerebral and cerebellar cortex, subcortical white matter, basal ganglia, brain stem and upper spinal cord.

MATERIAL SUBMITTED: Area of calcarine cortex and subcortical white matter.

Points for Discussion:

1. Diagnosis (one or more)

2. Nature of reactive changes