CASE 2

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Clinical abstract: The patient was a 22 year old male who presented 13 months prior to death with a two month history of nausea, vomiting, headache, transient right-sided weakness and intermittent diplopia. He had emigrated to the United States at the age of 19, and denied homosexuality or intravenous drug abuse. On admission to Queens Hospital Center his neurological examination revealed upper and lower extremity tremors and right lateral gaze nystagmus. Analysis of the CSF revealed 184 WBC/μl with 98% mononuclear cells and 2% polymorphonuclear cells, 310 RBC/ul, glucose 11 mg/dl, protein 217 mg/dl, inflammatory cytology, and negative results on VDRL, latex fixation for cryptococcal antigen and counterimmunoelectrophoreisis for meningococcus, Haemophilus influenza and pneumococcus. Head computerized tomography (CT) demonstrated hypodense lesions with mild contrast enhancement in the left frontal lobe and at the genu of the left internal capsule, consistent with subacute infarcts. The patient was placed on INH, Rifampin and ethambutol.

The patient returned 8 months later with complaints of frontal headache, blurred vision, weakness, fatigue, slow deterioration in cognitive abilities, and occasional urinary incontinence. On admission he was oriented to person and place, and had slowed speech and impaired short term memory. He displayed bilateral frontal release signs and was diffusely hyperreflexic and ataxic. Lumbar puncture yielded 79 WBC/ul with 82% lymphocytes, 16% polymorphonuclear cells and 2% monocytes, 12 RBC/ul, glucose 18 mg/dl, and protein 208 mg/dl. Head CT showed the previously noted areas of infarction and a new hypodensity in the left parietal lobe. The patient underwent a cerebral angiogram, which demonstrated multiple areas of stenosis and occlusion in all branches of the Circle of Willis and hypertrophy of collateral vessels at the base of the brain in a "puff of smoke" appearance. One month after admission the patient developed generalized tonic-clonic seizures. Over the remaining 4 month hospital course he experienced a gradual decline in cognitive function, punctuated by intermittent seizures. Two days before death the patient developed a Pseudomonas aeruginosa urinary tract infection. He subsequently became lethargic, developed clinical sepsis, and died.

At autopsy, the base of the brain was covered by thickened, fibrotic leptomeninges. Arteries of the Circle of Willis were markedly narrow, firm, grey and nodular to palpation. On section, multiple areas of recent and old necrosis were present throughout the cerebral and cerebellar hemispheres.

Material submitted: H & E and unstained slides from basal ganglia or cerebral cortex

Points for discussion: Diagnosis