CASE 1990-1

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Case Reference # 85-88

Clinical History: A 2 2/12 year old black male was admitted to CHOP because of developmental regression followed by the sudden onset of focal seizures, left hemiparesis and fever.

He was born at term following an uncomplicated pregnancy and delivery, but a tracheo-esophageal fistula, patent ductus arteriosus and coarctation of the aorta were diagnosed. The fistula and cardio-vascular defects were repaired and a gastrostomy was done. The tracheo-esophageal fistula repair broke down several times over the next 18 months, eventually necessitating a colonic interposition and Nissen fundoplication.

Examination showed a rectal temperature of 38.7°C and extreme irritability. Pupils were equal and reacted to light. There was a conjugate right gaze preference but EOMS were complete with oculocephalic maneuver. He did not respond to visual threat nor did he attend to verbal cues. The neck was supple, he moved all extremities semipurposefully and tone was diffusely increased. DTRs were 2/4 throughout and plantar responses were extensor. A draining fistula was present on the left anterior abdominal wall.

Diagnostic studies: WBC 10,000/mm^3 with 67% segs, 23% lymphs, 7% monos and 3% eosinophils; Hgb - 9.9 gm/d; platelets-593,000/mm^3; CSF - 864 WBC/mm^3 (81% segs and 19% lymphs), 3040 RBCs/mm^3, glucose 54 mg/dl and protein over 600 mg/dl. Spinal fluid cultures for bacteria, fungi, mycobacteria and viruses were negative. A CAT scan revealed moderate ventricular dilatation, right frontal lobe enhancement and multiple areas of cerebral infarction of variable ages.

Comprehensive immunologic evaluation yielded no abnormalities, but skin test for Candida antigen was positive.

Infant was treated with Oxacillin, chloramphenicol and Acyclovir, but continued to deteriorate, and expired 3-4 months after onset of symptoms.

Necropsy Findings: Large meningeal mass on ventral surface of brain stem (see kodachrome), smaller nodules at carotid bifurcation and over cervical and lumbar cord.

Material Submitted: Kodachrome of ventral surface of brain, one H&E and one unstained section of meningeal masses.

Points for discussion:

1. Diagnosis
2. Pathogenesis

Fungus type?