

Case 4

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Clinical Abstract:

The patient was a 34 year old female former IV drug abuser who was born with Ebstein's anomaly. At age 9 she underwent tricuspid valve plication, atrial septal defect repair and pacemaker insertion. She presented in July with right sided weakness and paresthesias, numbness of teeth, gums and tongue, auditory hallucinations, paranoid ideation and difficulty speaking and writing. A neurology consultant found no reason to suspect an organic brain syndrome, so she was admitted to the psychiatry service for three months. EEG, while on low dose Haldol, revealed diffuse slowing. A Bender test was stated to be compatible with an organic process. CSF, in July, contained 9 WBCs (predominantly mononuclear) and had a protein of 32 (24% gamma globulin). Six weeks later the CSF had a single WBC and a protein of 32.

She was readmitted the following March for two months complaining of inability to function, increased agitation and auditory hallucinations. CT of the head revealed no abnormalities. A switch from Haldol to Stelazine coincided with improvement in her condition.

Her final admission began in July and ended with her death in September. On admission, she was depressed with a flat affect and psychomotor retardation. Examination revealed disorientation, poor memory, anomia, stiffness, hyperreflexia of the lower extremities and frontal release signs. In the initial hospital course there were episodes of unresponsiveness with drooling and diaphoresis that lasted several minutes. CSF studies (9/82) revealed 35 WBCs (mostly mononuclear), a protein of 35 and equivocal oligoclonal banding. Serum measles antibody titers were 1:40. She suddenly became febrile (104.5°) and minimally responsive with decreased reflexes and incontinence. No source for the fever was found. The next day she had a generalized seizure and died later that day with a cardiac arrhythmia.

Necropsy findings included surgically repaired congenital heart disease, dilatation of the atria and the right ventricle, biventricular hypertrophy, acute and chronic myocarditis, chronic persistent hepatitis with fibrosis and hemorrhagic cystitis. The brain weighed 1240 gms. Gross examination of the brain and spinal cord was unremarkable.

Material submitted: One LFB-PAS stained section of the pons.

Point for discussion: How would you classify this condition?