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Case 2001-05.

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Clinical history:

A young male was seen for the first time at the age of 1 0 with symptoms of intracranial hypertension. Previous history indicated similar symptoms since the age of 7.A CT showed communicating hydrocephalus, and a ventriculoperitonial shunt was placed. CSF analysis at this time showed 3.5cells/mm3, protein 10mg/dl, glucose 67 mg/dl. In the following 18 to 24 months, the patient was admitted three more times due to shunt blockage needing valve replacement. During this time, he had two CSF revealing 53 and 32 cells, protein 18 and 100, glucose 67 and 12, respectively.

Eighteen months later he sought medical attention due to inability to control anal sphincter for the past 3 months, dysfunction of bladder sphincter in the past 30 days, acompannied by muscle weakness, impotence, weight loss and fever in the previous month. Patient was admitted for evaluation, which revealed hepatosplenomegaly, weight loss and leucopenia in peripheral blood . Chest x-ray showed diffuse micronodules, a calcified granuloma in the RLL and hilar calcifications. Search for ova in feces was negative. Screening for Rubella and CMV was positive for IgG only, and negative for blastomycosis, toxo, HEP B , HIV and TB. A definite etiology for the process could not be found and patient was discharged a month later .

Patient was readmitted two months later because of constipation and weakness in lower limbs for the past 7 days. Physical examination revealed marked weight loss, hepatomegaly, left hemiparesis and decreased muscle strength in lower limbs, which progressed to further paralysis with enuresis. CSF analysis showed 53 cells/mm3, protein 4026 mg% and glucose 25mg%. CSF cultures were negative.

He was started on steroids, with improvement of both muscle strength and control of anal sphincter, and able to walk with help. Another CSF analysis showed 0.7 cells/mm3, protein 5 1 50mg%, glucose 25mg%. CSF imunology for schistosomiasis, syphilis, cysticercosis and toxo was negative. A rectal biopsy in search of ova was negative.Steroid therapy was terminated one month later, followed by worsening of neurological symptoms. An MRI of his spinal cord at this time was c/w myelitis. Reintroduction of steroids was ineffective.

The following month, patient developed respiratory distress, muscle weakness in upper limbs, and hypoesthesia at T6 level. A chest CT showed diffuse micronodularity, s/o active inflamatory process in the lung. Patient died one and a half months later due to sepsis and respiratory failure.

Necropsy findings included diffuse thickening of dura mater, thickened leptomeninges on the ventral surface of brainstem at the level of VI, IX, X, XI cranial nerves; communicating hydrocephaly; duplication and tearing of septum pellucidum. The dura mater was adherent to the leptomeninges in the entire spinal cord; the leptomeninges were dusky, thickened and adherent to the spinal cord from C3-C4 to cauda equina. Sectioning revealed asymmetric lesions involving all levels of spinal cord.

Material submitted: H&E of section of spinal cord
Unstained section of spinal cord

Points for discussion: 1. Diagnosis
2. Pathogenesis