

CASE 7

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Clinical Abstract:

This 16 year old boy developed fever, irritability, and papular skin lesions at 4 months; these lesions became scaly and developed clear centers. Simultaneously, generalized lymphadenopathy and hepatomegaly were noticed. Headaches and minor seizures began at 4 years of age. The skin rash recurred intermittently and improved with Prednisone. One skin biopsy showed "vasculitis". Infectious and immunologic work-up was unrevealing. He regressed neurologically and intellectually and at age 10 his mental age was 5 years. Generalized seizures, refractory to treatment, appeared. At 13 years of age, ventricular enlargement required insertion of ventriculo-peritoneal shunt. The CSF opening pressure was 230 mmH<sub>2</sub>O and CSF contained 62 WBC's with 23% lymphocytes and 76% polymorphonuclear leukocytes, 47 mg/dl sugar and 208 mg/dl protein. By 15½ years he could no longer walk or sit unsupported and was unable to speak or understand spoken language. There was flexion posturing of all four extremities. Multiple small regions of low density were seen in both hemispheres on CT scan. He died at age 16.

Necropsy Findings:

Fibrous scars were present in the liver, kidneys and spleen. Skin lesions could not be identified and random sections did not show significant pathologic changes. Leptomeninges were thick and fibrotic and leptomeningeal vessels had a cord-like appearance. There were multiple lesions in the brain.

MATERIAL SUBMITTED: One H & E-LFB stained section from the right fronto-temporal operculum and insula, representative of many lesions.

Points for Discussion:

1. Diagnosis or differential diagnosis
2. Relationship between the visceral, skin and CNS lesions