

Case 5

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Clinical Abstract: This twenty year old woman first developed abdominal pain, sporadic vomiting, jaundice and elevated liver enzymes. She was treated for duct obstruction and pancreatitis.

She became debilitated and developed malabsorption of fat. Serum glucose was 870 mg/dl; Na⁺ 126 and K⁺ 7.5 mEq/l. She had episodes of diarrhea and fainting. Serum T4 was 2.4 mg/dl and TSH was 18.6 u/U/ml; antithyroid antibodies were detected, and synthroid was begun. ANA was strongly positive. Medical management was complicated by recurrent vomiting and intractable abdominal pain requiring narcotics. A choledochoduodenostomy, distal subtotal pancreatectomy and a pancreatico-jejunostomy were done. Postoperative serum osmolarity of 341 with urinary osmolarity of 164 indicative of diabetes insipidus; pitressin was begun.

Cerebral CT scan to evaluate diabetes insipidus showed multiple contrast-enhancing lesions in both cerebral hemispheres, most concentrated in the corpora striata. She displayed subtle behavioral changes but no hard neurologic signs. Spinal tap on 5/23 showed slightly elevated protein but was otherwise normal; cultures were negative. Hydrocortisone was begun because of repeated low cortisol levels. HIV and toxoplasma titers, and PPD were negative. She then became afebrile, abdominal pain diminished, mental status improved and she began eating a regular diet. However, there was a significant progression of size and number of the enhancing lesions in both cerebral hemispheres. She had a brain biopsy.

Material Submitted: Brain biopsy.

Points for Discussion:

1. Diagnosis
2. Pathogenesis
3. Treatment