

CASE 1994-6

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Clinical History:

The patient was a young female with a history of movement disorder following a viral syndrome at the age of 21 years, when she started having excessive drooling and involuntary lip smacking. Later on she developed involuntary tongue movements, tongue biting, difficulty with articulation and involuntary movement of her limbs and occasional dystonic posturing. She also had strange bizarre speech and behavior and tendency to unintentionally injure herself. At age 28, she had severe choreiform movements of arms and lips. Since she was not able to take care of herself, she was admitted to a nursing home. She continued to deteriorate with increased athetoid movements and self destructive behavior with chewing and biting her cheeks and died at the age of 31.

Past History: Infectious mononucleosis at age 20.
Family History: Grandmother had oral involuntary movements in later years.
Physical Exam: Thin white female, emotionally labile, showing lip smacking, drooling and repetitive movements of lips, dysarthric speech, unable to stand.

Neurological

Exam: Cranial nerves - normal, no nystagmus
Motor - mild decrease of tone, no focal weakness, reflexes-hypoactive diffusely.
Sensory exam - normal
Mental - IQ 106

Labs: Chest x-ray normal, EKG - normal
Blood Glucose 100, LDH 268, Cholesterol 176
Copper and ceruloplasmin (serum and urine) - normal
CSF - normal

Necropsy Findings:

Degeneration of basal ganglia

Material Submitted:

1. Kodachrome of corpus striatum (gross)
2. H&E stained section of corpus striatum

Points for Discussion:

1. Differential Diagnosis
2. Plan for further workup.