

## 52nd ANNUAL DIAGNOSTIC SLIDE SESSION 2011

### CASE 2011-1

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**Clinical History:** The patient was a 56 year-old female with a past medical history of hypertension, hypercholesterolemia, and insulin-independent diabetes mellitus who presented with a five month history of diminished facial expression and voice softening. A modest cognitive deficit was also noted. Thus, a diagnosis of Parkinson Disease with accompanying dementia was given and Sinemet therapy was initiated. Her initial brain MRI at that time showed abnormal signal within the left basal ganglia and bilateral changes in the periventricular white matter (Images enclosed).

Despite the therapy, her neurologic status slowly declined. Two months after her initial workup, she was admitted for severe metabolic acidosis due to metformin toxicity; by this time she had increased memory loss and disorientation. Her gait had also worsened to the point where she needed a walker for ambulation.

Three months after the initial presentation, she was hospitalized again with worsening speech slurring and right-sided weakness. An MRI showed multiple areas of abnormal T2 hyperintensity in the bilateral cerebral hemispheres, midbrain, and pons. Additional foci of T1 hyperintensity were noted in the right caudate nucleus, as was a nonspecific area of enhancement in the posterior right frontal lobe. (Images, without enhancement, enclosed). A lumbar puncture showed only mildly elevated protein. The right frontal lobe was biopsied and showed vascular fibrinoid degeneration, intramural/perivascular lymphocytic infiltrate, occasional fibrin thrombi, and perivascular microhemorrhages most consistent with vasculitis (not shown). She was started on steroids and Cellcept, but continued to decline until her death one year after her initial symptoms began.

Key autopsy findings included extensive bilateral and symmetrical necrosis of the basal ganglia (see gross images A and B) and substantia nigra (gross image C).

**Material Submitted:**

1. H&E stained slide of left basal ganglia/thalamus
2. Serial MRI imaging
3. Postmortem gross photographs.

**Points for Discussion:** Additional stains, differential diagnosis, review of literature