54th ANNUAL DIAGNOSTIC SLIDE SESSION 2013

CASE 2013-2

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Clinical History: A 69 year old woman with history of hypertension and hypothyroidism developed neck pain and stiffness accompanied by severe headache two days after a suction-assisted lipectomy of her anterior neck. She received Cephalexin before and after surgery. She went to her local E.R. where her temperature was 99F. She had mild ecchymoses and tissue swelling related to surgery but no evidence of wound infection. There were no neurological symptoms. A lumbar puncture and a CT scan of her head were normal. She received hydromorphone (1.5mg, I.V.) and diphenhydramine (25mg, I.V) and was admitted to the hospital. Overnight she received a further 1mg hydromorphone, I.V. and 25mg diphenhydramine, I.V. She "slept throughout the night" but the following morning was unresponsive with a blood pressure 80/40mmHg and oxygen saturation of 60% on room air. She was treated with Narcan and discharged home, almost at baseline, three days later.

Three weeks later she drove her daughter's car to a relative's house, not realizing the car was not her own. She was later found attempting to put her panty hose on over her jeans. She was brought to a local E.R. and had multiple mild memory and attention deficits. Over the following 72 hours she had a rapid profound decline in mental status, could not remember family members and stated she had 100 children. She forgot aspects of her recent hospitalization and became fearful, uncooperative and less interactive developing urinary incontinence, facial tics and repetitive nose-picking. On examination she was awake but non-verbal and not following commands. She was intermittently agitated, with startle myoclonus. She moved all limbs symmetrically and had flexor plantar responses. An extensive laboratory work-up including CBC, chemistries, liver function, amylase/lipase, ammonia, B12, B1, thyroid function, lactate, lyme, RPR, HIV, WNV, EEE, Ehrlichia, Babesia, Anaplasma, serum and urine heavy metals, paraneoplastic screen and CSF 14-3-3 were negative. Neuroimaging showed diffuse confluent T2/FLAIR white matter signal hyperintensity.

She remained mute, became progressively rigid and akinetic, and was transferred to a neurorehabilitation facility. Three months later she was alert and responsive, could answer yes/no questions and follow short simple commands. By 6 months she could follow all commands, rapidly name the days of the week backwards and had fluent speech. She described in detail how her back pain was alleviated by removal of an 80 foot and a 30 foot snake from her abdomen. At 9 months she resided in an assisted facility and was increasingly suspicious, paranoid and delusional. She was inattentive and disorientated, answered questions tangentially, and showed impairments in memory encoding, executive function, behavioral control, and visual spatial function. She died approximately 2 years after her first presentation.

Autopsy: The brain weighed 1300gm and coronal sections revealed widespread white matter discoloration and softening with ventriculomegaly.

Material submitted: Stained (Luxol Fast Blue, H&E) and unstained section of left frontal cortex

Points for discussion:
1. Diagnosis
2. Pathogenesis