CASE 2014-10

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Clinical History:
A 76 year old man with hypertension and hyperlipidemia presented with 1 week of painful right T7 dermatomal rash and paresthesia in the bilateral lower extremities. Physical exam noted right T7 vesicular/erythematous rash, full strength throughout with the exception of mild hip flexor weakness. Reflexes were absent at the knee and trace at the ankles. He walked with an ataxic gait, had diminished pain, light touch and proprioception to the knees. He was treated with Solumedrol and Acyclovir IV. Thoracic spine MRI was limited by patient discomfort. Within several hours, he had speech difficulty, and right arm and bilateral leg weakness. Exam revealed intact cranial nerves, upper limb strength but 0/5 hip flexor strength bilaterally. The following day, he developed dysphagia and had increased work of breathing. He now had 0/5 strength throughout the upper and lower extremities with the exception of 4/5 strength in the hands and ankles. Reflexes were absent.

CSF glucose 62, total protein 191.5, RBC 11857, WBC 130 (80% Neut, 16% Lymph); Serum varicella zoster virus PCR was positive; CSF positive IgG to VZV.

He was intubated after which he was awake following commands. Eyes were open with full extraocular movements. He had profound proximal greater than distal weakness in all extremities. His exam deteriorated and by day 6 he was ophthalmoplegic and quadriparetic. His blood pressure was labile on pressors. Spine MRI showed questionable cauda equina enhancement. Brain MRI showed enhancement of cranial nerves III and the right VI/VIII nerves. EMG/NCS revealed severe motor and sensory axonal polyneuropathy with demyelinating features. On day 17, repeat brain MRI was abnormal (see image). After family discussion, he was made comfortable and extubated. An autopsy was performed.

Autopsy findings:
Zoster-like skin ulcers, no evidence of ongoing/active VZV skin infection (no viral cytopathic changes; negative VZV immunostain)
Hepatosplenomegaly
Pulmonary edema

Material submitted:
H&E section(2) right frontal lobe and cauda equine; MRI brain T1W post contrast, Coronal section at genu of CC.

Points for discussion:
Is there a unifying diagnosis?