60th ANNUAL DIAGNOSTIC SLIDE SESSION 2019

CASE 2019 [10]

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Clinical History:
A middle-aged man from rural western US with a history of an NSTEMI, reflux, hyperlipidemia, hypertension, depression and Poland syndrome presented by ambulance to the ER for wheezing and hyperventilation. He had visited the ER several times over the previous 5 days for a reported industrial work place accident for which he received dexamethasone. In the ER, he continued to complain of severe muscle spasms and paresthesias in his neck, back and right arm, and reported that he had also been unable drink fluids due to these spasms. Physical exam showed him to be afebrile, mildly dehydrated and tachypneic with good oxygen saturation. After admission, he became acutely delirious prompting transfer to a higher acuity hospital. Upon arrival he demonstrated florid psychosis, dysphasia, dystonic posturing, akathisia and orofacial dyskinesias. Electrolytes were normal but his Cr, BUN and CPK were elevated. Following intubation and sedation, brain and C-spine MRIs and a lumbar puncture were normal. An EEG showed generalized slowing and disorganization consistent with encephalopathy. During repeat interviews with his family, they also reported a witnessed ground level fall, possible auditory hallucinations, travel history to the Pacific Northwest and a grouse hunt. By day 3 of admission his blood pressures began fluctuating and he developed a temperature of 40°C. Over the following 48 hours his condition deteriorated and he became unresponsive with EEG showing status epilepticus and bitemporal lobe edema on MRI. On day 10, his MRI showed diffuse edema in the pons and midbrain, bilateral thalamic and temporal pole hyperintensities with worsening leptomeningeal enhancement and corticospinal tract and cranial nerve enhancement in CN III and V.

His blood and CSF cultures as well as PCR and serology tests were negative for bacteria, viruses, fungi, syphilis, *Francisella*, *Borrelia*, *Leptospira*, *Ehrlichia* and typhoid. Vitamin levels, thyroid hormones, toxicology and drug studies were also normal. An ANA was 1:160 but all other autoimmune and demyelinating evaluations were negative.

Given his poor prognosis, he was placed on comfort care and passed away on hospital day 15.

Autopsy findings:
The autopsy was remarkable for cerebral edema but no other significant gross pathology. Pending studies at the time of death included a 14-3-3/tau and tests performed by the CDC.

Material submitted: H&E section of cerebellum.

Points for discussion:
1. Explain the differential diagnosis for rapidly progressing encephalopathy.
2. Ancillary studies.