61st ANNUAL DIAGNOSTIC SLIDE SESSION 2020.

CASE 2020 [#11]

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Clinical History:

The patient was a 56-year old man presented with confusion. Ten months prior to presentation he developed fatigue, joint pains, and weight loss. Serologic testing for Lyme disease was negative but he nevertheless received a 3-week course of doxycycline without improvement. He was diagnosed with fibromyalgia by his local physicians. Three months prior to presentation he developed insomnia and cognitive difficulty which led him to quit his work as a nanotechnology engineer. During the month prior to presentation, he frequently became lost while walking and had difficulty spelling words and dressing himself. His past medical history was notable for mantle cell lymphoma diagnosed nine years prior which was treated with bendamustine and in complete remission on maintenance rituximab. His family history was notable for schizophrenia in his sister and mild cognitive impairment in his mother. Initial examination showed disorientation to time, slow sparse speech, and poor recall with psychomotor slowing and masked facies.

Three weeks later, his symptoms worsened prompting presentation to the ED. On examination, he was afebrile with normal vitals but was disoriented to time and place and exhibited slow sparse speech with poor repetition, writing, and fluency, difficulty following complex commands, left-right confusion, and difficulty performing simple arithmetic. Cranial nerve examination was normal, and he was full strength with normal tone, no tremor, asterixis, or bradykinesia. Reflexes and cerebellar testing were normal, and toes were downgoing. There was a non-engorged tick in his left groin with surrounding induration but no rash. Multiple lumbar puncture CSF studies showed pleocytosis but unremarkable flow cytometry and cytology. RT-QuIC was negative. Broad auto-immune and infectious serologic testing was unrevealing. Magnetic resonance imaging showed no specific findings. Empiric solumedrol did not improve his symptoms.

He was discharged after an 11-day hospitalization but presented again to the ED two days post-discharge with worsening agitation. He was admitted, and olanzepine and thorazine were initiated to control his symptoms. He continued to decline and one week later he was unable to name any simple objects or follow any commands. His alertness deteriorated in the following week and he was only able to mumble incomprehensibly. Tone was diffusely increased with brisk reflexes and crossed adduction. Toes were downgoing. He remained full-strength and ambulatory to his bathroom with assistance from nursing. After another week, he had no further verbal output, would open his eyes to loud stimulation but not regard or track, and developed decorticate posturing. IVIG and steroid treatments did not improve his symptoms. He was transitioned to comfort care, and he passed away ten weeks after his initial presentation.

Autopsy findings:

The fresh brain weight was 1240 grams. There were no significant gross neuropathologic findings.

Material submitted:

1.) H&E stained glass slide of the right insula
2.) Whole slide scans of H&E stained sections of the right insula, left frontal lobe cerebral cortex, and cerebellum

Points for discussion:

1.) Differential diagnosis and work-up. 2.Epidemiology and pathology