Encephalopathy Secondary to Aggressive CD5+ DLBCL Versus Herpes Zoster Encephalitis.

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Published In/Presented At

Kass, K. (2017, October 28). *Encephalopathy Secondary to Aggressive CD5+ DLBCL Versus Herpes Zoster Encephalitis*. Poster Presented at: American College of Physicians (ACP), Harrisburg, PA.

Encephalopathy Secondary to Aggressive CD5+ DLBCL Versus Herpes Zoster Encephalitis

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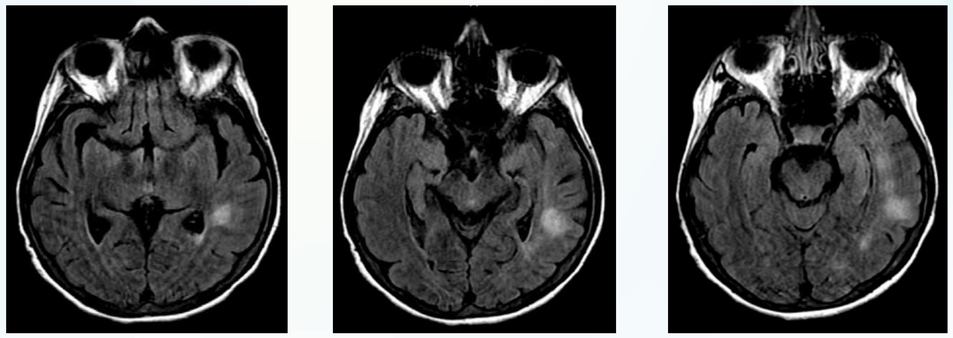
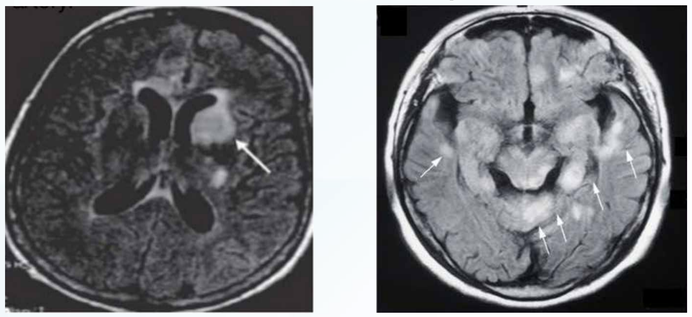
INTRODUCTION

Diffuse large B-cell lymphomas (DLBCL) positive for immunohistochemical marker CD5 are uncommon, non-Hodgkin lymphomas that can rarely present with profound encephalopathy and offer affected patients unfortunately poor prognoses. Deterioration within two weeks though is unlikely. The author postulates the possibility of a subacute, untreated varicella zoster infection as the etiology of a PCR-negative zoster encephalitis that precipitated this patient's decline due to incidental immunocompromise.

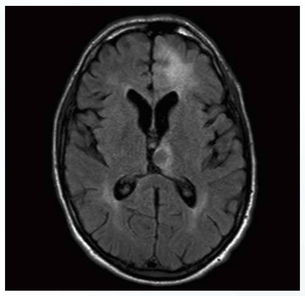
MRI FLAIR of this Patient on Presentation



MRI FLAIR of VZV Encephalitis



MRI of Classic DLBCL (Primary CNS Involvement)



CASE REPORT

A 66-year-old female with a history of hypothyroidism and subacute resolution of an untreated herpes zoster infection[1] presented with a one month history of encephalopathy, lightheadedness, nighttime fevers and fatigue. She was febrile, hypotensive and tachycardic, with pallor and a mildly tender abdomen. Mental status exam revealed disorientation, inconsistent command following, inattention, perseveration, confabulations and 6/30 on the Montreal Cognitive Assessment. Imaging demonstrated splenomegaly and mesenteric lymphadenopathy concerning for lymphoma. Brain MRI revealed T2 hyperintensity in the left temporal lobe with suspected internal hemorrhage on susceptibility weighted imaging. Lab studies showed pancytopenia, elevated inflammatory markers, and significantly elevated LDH. CSF studies revealed elevated protein and RBCs with normal glucose, though PCR for VZV was negative. CSF PCR sensitivity for varicella is only 80%. Cytology on CSF was negative, but this test has an even lower sensitivity. Paraneoplastic antibodies were checked and negative. By day 11 the patient became hemodynamically unstable and developed multiple organ failure. Exploratory laparotomy performed for suspected ischemic colitis instead revealed bulky mesenteric lymphadenopathy obstructing the small bowel, which was resected and biopsied. The patient further deteriorated and the family withdrew care soon after. Lymph node biopsy revealed aggressive CD5+ DLBCL, activated-B-cell-like. Autopsy and brain biopsy were not performed.

1. The patient visited her PCP 3 months prior to her presentation to the hospital, complaining of LLQ abdominal pain of one month duration. Uncomplicated shingles was diagnosed, of one dermatome, and Valacyclovir was prescribed though never taken. She elected to treat symptoms with Neurontin and Aleve. She was seen by her PCP for follow up 6 weeks later with near resolution of the shingles infection.

DISCUSSION

We do not know what provoked this patient's multi-organ failure and rapid deterioration. DLBCLs rarely present with encephalopathy and CD5+ DLBCLs, though notoriously aggressive, do not by themselves precipitate such rapid deterioration. Herpes zoster encephalitis can cause rapid decline but is a rare diagnosis in the presence of "sterile" CSF and unilateral temporal lobe findings. The author suspects CD5+ DLBCL caused immune compromise such that herpes zoster encephalitis instigated a rapidly fatal outcome in this patient. The case illustrates rare presentations of two coinciding diseases. Additionally it emphasizes the paramount need for a thorough history of pre-admission events and for broad, empiric coverage of patients even in the face of negative or uncharacteristic evidence. Yes, this patient was found, post-mortem, to have a famously aggressive DLBCL of the abdomen, but she presented with encephalopathy. And, when all of the pieces do not fit the clinical presentation, other factors need to be considered. Given the presentation and the above date there is room to believe that aggressive antiviral treatment may have changed the clinical course of this patient.