Case Report
CNS Intravascular Lymphoma: A Case Report

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Intravascular lymphoma is a rare but well-described entity. The clinical manifestations are heterogeneous. We report a case of a 59-year-old woman who presented initially with syncope followed by subacute cognitive decline that progressed to minimally conscious state. Shortly after the transfer to our tertiary center the patient died. Brain autopsy disclosed the diagnosis of B-cell intravascular lymphoma. We speculate that syncope could be the first manifestation of central nervous system intravascular lymphoma and should be considered in the differential diagnosis of unexplained syncope. In addition, we stress the importance of early brain biopsy in unexplained white matter disease.

1. Case Report

The patient was a fifty-nine-year-old right-handed school counselor Caucasian woman with a past medical history remarkable for left-sided breast cancer, in remission, status postresection ten years prior to admission, hypothyroidism on thyroxin, and hyperlipidemia on rosuvastatin. She was transferred to our neurology tertiary center, from a local hospital, for evaluation and treatment of unexplained altered mental status. The patient’s history dates back to four months when she suffered an episode of loss of consciousness consistent with syncope. She suffered prodrome lightheadedness. Shortly after that, everything turned black and then blacked out for few minutes. During the episodes she was able to hear people talking to her but could not respond. No jerking was witnessed. In addition, there was no tongue biting or urinary incontinence. She woke up clear-headed with complete recollection of the event. The event last for about 60 seconds. At that time, she underwent cardiac evaluation which was unremarkable.

About two months after the syncopal episode, the patient’s family and coworkers started noticing intermittent problems with her cognitive function. Specifically, she displayed difficulties using her computer. However, she continued to work but had progressive difficulties with common daily tasks. One month later, the patient was admitted to a local hospital with an acute and transient episode of dysarthria and facial palsy. A brain magnetic resonance imaging (MRI) was completed to exclude a cerebrovascular etiology of her acute symptoms. The MRI was reported as abnormal due to the presence of “white matter disease”. While in the hospital, she became progressively worse and her mental status deteriorated quickly over the course of one week. Repeat brain MRI was reported as “significantly worse” than the initial one and showed multiple areas of white matter lesions some of which showed restricted diffusion. Cerebrospinal fluid analysis was pursued to exclude intracranial infections and the only abnormality reported was a high protein level. The exact number was not available. Full anticoagulation with unfractionated heparin and coumadin was started as well as empirical treatment with acyclovir. Acyclovir was discontinued after a viral infection was excluded by cerebrospinal fluid (CSF) polymerase chain reaction (PCR) studies. The patient continued to deteriorate over the next week to a minimally responsive state. No details were available from the records we reviewed on her predeterioration status. A computerized tomography (CT) of the head disclosed progression of the white matter disease with interval development of a left parietal parenchymal...
hemorrhage, in addition to diffuse cerebral edema. Anticoagulation was reversed, and she was transferred to our hospital for higher level of care.

Upon admission to our institution the patient was in a minimally conscious state, which allowed only a limited neurological examination. She was arousable to voice and followed one-step commands inconsistently. She was able to repeat and answer only simple questions inconsistently as well. Pupils were equal and reactive. Corneal reflexes were present bilaterally. Visual fields were intact to threat. No gross restriction of extraocular muscle movement was noticed. No gaze deviation of skew deviation was noted. No nystagmus was observed. Her face was symmetric. The tongue was midline with no atrophy or fasciulations. Motor examination was consistent with a mild-moderate spastic pyramidal left hemiparesis. No involuntary movements were seen. Muscle stretch reflexes were brisk (3+) bilaterally. Plantar reflexes were extensor bilaterally. The patient was able to localize tactile and noxious stimuli on both sides without obvious side-to-side difference.

A complete blood count was remarkable for anemia and thrombocytopenia (hemoglobin 9.5 mg/dL and platelet count of 83,000/mm³ respectively). Results of a basic metabolic panel, which included serum electrolytes, blood glucose, and kidney-function tests, were within normal limits. Liver function tests revealed elevated liver enzymes: alanine aminotransferase 82 IU/L, aspartate aminotransferase 96 IU/L; albumin low 2.8 mg/L; ammonia elevated to 57 mg/L. Inflammatory markers were abnormal: erythrocyte sedimentation rate 36 and C-reactive protein 1.9. Antinuclear antibody was positive with a low titer of 1:320. Other pertinent normal/negative studies: coombs, platelet antibodies, heparin-induced thrombocytopenia antibodies, lupus anticoagulant antibodies, antiphospholipid antibodies, hepatitis C and B, copper, vitamin B12, homocysteine, anticardiolipin antibodies, antithrombin III, factor V Leiden, protein C, protein S, lipoprotein A, prothrombin G20210A, antithrombin, lupus anticoagulant antibodies, antiphospholipid antibodies, heparin-induced thrombocytopenia antibodies, and antithrombin. Other tests included: leukemia panel FISH (normal), SMA 12 (typical pattern), and viral panel (negative).

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Figure 1: Multiple confluent white matter lesions demonstrated by 3 mm axial fluid attenuated inversion recovery (FLAIR) brain magnetic resonance images (MRIs) demonstrating and diffusion weighted imaging (DWI), respectively.

Figure 2: Brain biopsy demonstrating the presence of intravascular atypical lymphocytes.

Intravascular lymphoma is a rare and fatal disease that presents with a myriad of nonspecific symptoms. To ascertain the earliest diagnosis possible, and to potentially alter patients’ outcomes, we propose to include intravascular lymphoma it as a differential diagnosis in patients presenting with syncope of unclear etiology. Brain MRI should be considered in evaluating patients with unexplained syncope. Finally, the cases raise the importance of early brain biopsy in unexplained white matter disease which could positively impact the course of a progressive fatal disease.

Conflict of Interests

The authors declare no financial disclosures or conflict of interests.

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