Case Report

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Intravascular Lymphoma Mimicking Acute Haemorrhagic Leucoencephalitis

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Abstract

Intravascular lymphoma and Acute Haemorrhagic Leucoencephalitis are rapidly progressive diseases with poor prognosis. Differentiated between CNS variant of IVL and AHLE can be challenging, as they both can be clinically identical. Our case is about 63-year-old female, who presented with rapid onset of cognitive impairment. During her hospitalization, another rapid deterioration has occurred, and she became tetraplegic, with aphasia, and obtunded. Brain MRI showed many confluent lesions in the white matter, with. Most lesions appeared hemorrhagic. After the MRI, a radiological and clinical differential diagnosis between intravascular lymphoma and Acute Haemorrhagic Leucoencephalitis was made. Rapid brain biopsy has revealed the diagnosis of IVL. She had a good responsive to therapy, and regained most of her cognitive and motor functions.

Keywords: Intra-vascular lymphoma; Acute haemorrhagic leucoencephalitis

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Introduction

Intravascular Lymphoma (IVL) is a rare lymphoma, most commonly of B-cell origin. It is classified according to epidemiology and organ involvement: patients of Western variant present frequently with CNS and skin involvement, while in those of Asian variant bone marrow, spleen and liver involvement is more common [1]. Clinical presentation depends on the organ involved and is related to vascular occlusions. The diagnosis is made by identifying lymphoma cells within small to medium blood vessels from cutaneous biopsy or tissue from other organs, including the brain [2]. Acute Haemorrhagic Leucoencephalitis (AHLE) is a rare disorder characterized by an acute, rapidly progressive, monophasic, fulminant inflammatory haemorrhagic demyelination of white matter. Known triggers, such as infections, bone marrow transplantation, and complement mutation have been described and MRI imaging shows multiple focal white-matter lesions with hypointense central spots and bleeding [3].

Case Report

A 63-Year-old female, with a past medical history of hypertension and smoking, complained of two weeks of general weakness, myalgias, and subfebrile temperature. Her family reports changes in behavior including apathy and dressing inappropriately. Following an event of loss of consciousness, a non-contrast brain CT revealed chronic ischemic changes and bi-lateral hypodense lesions in the frontal lobes **(Figures 1A and 1B)**. On examination the patient was alert but disoriented and anomic, executes partial commands without any other focal neurological signs. No lymphadenopathy or hepatosplenomegaly were detected.

CBC showed mild anemia (Hb 10.6 g/dl), WBC, PLT and INR were within normal limits, as were the liver and kidney function. LDH was elevated at 1256 IU/ml (normal up to 480). CRP was 4 mg/dl. Repeated Lumbar punctures mild pleocytosis, but negative culture and PCR for infectious agents. A follow-up LP showed 15 MN cells and 8 PMN cells. Cytology from CSF was negative twice. Serology of wide infections screen was negative.

EEG showed an irregular and slow background, mostly in temporal regions. A total body CT showed few reactive lung nodules. Tran's esophageal echocardiogram was normal. Brain MRI **(Figures 1C-1H)** showed many confluent lesions in the white matter, with enhancement, no restriction on DWI and no gray matter involvement. In SWI most lesions appeared hemorrhagic. After the MRI, a radiological and clinical differential diagnosis between IVL and AHLE was made.

Clinically, four days after admission progressive deterioration occurred: The patient suffered severe global aphasia and tetraplegia. Due to her severe neurological deterioration, a single dose of corticosteroids was administered. A deep cutaneous

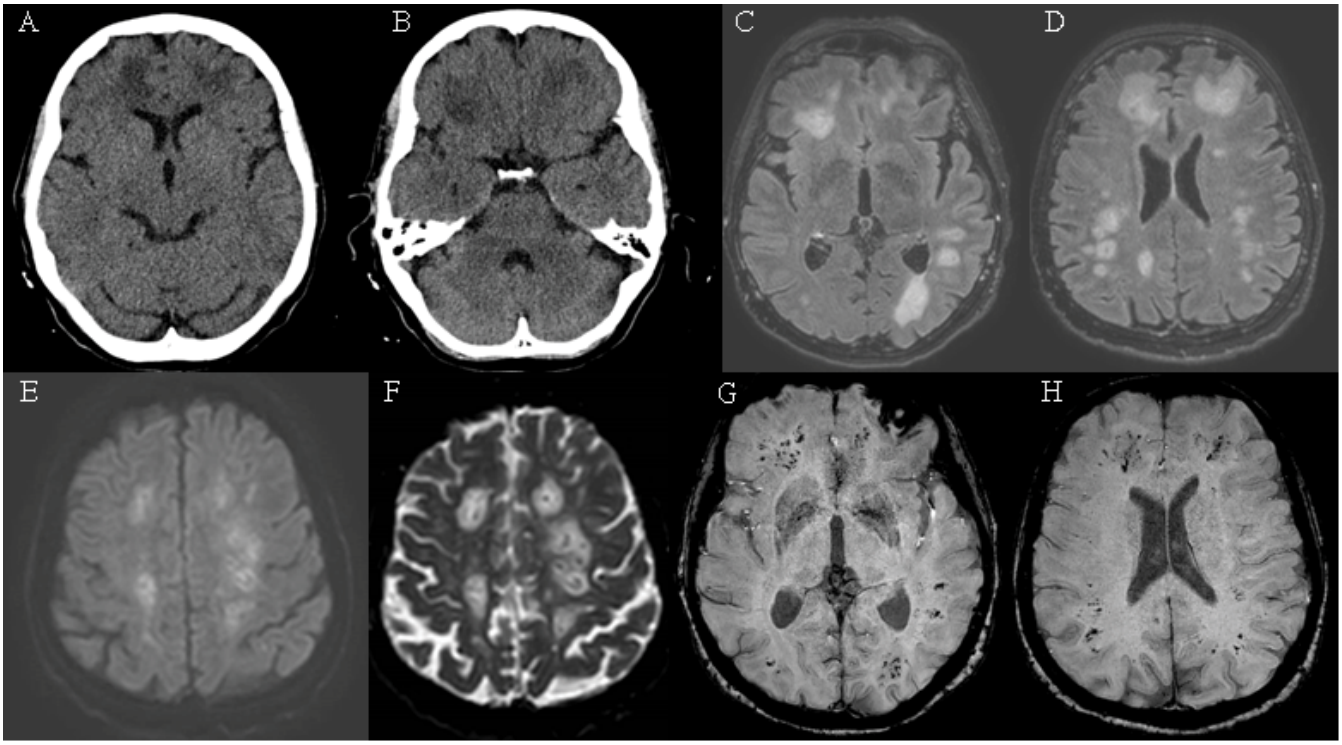

Figure 1 (A & B) Non-contract CT at arrival showing bi-frontal hypodense lesions. **(C & D):** Fluid attenuated inversion recovery axial MRI imaging showing bi-frontal and multi-focal increased signals. **(E & F):** Diffusion weighted imaging and apparent diffusion coefficient MRI imaging showing no restriction. **(G &H):** Susceptibility weighted imaging MRI imaging showing multifocal hemorrhagic lesions within white matter lesions.

biopsy was normal and brain biopsy was performed. Due to the rapid deterioration of the patient's mental status, and a suspicion of AHLE, steroid was initiated after the biopsy. The biopsy showed expansion of vascular cavities of large cell clusters in the vascular space, compatible with intravascular large B cell lymphoma with hemorrhages **(Figure 2)**, immunohistochemistry was positive for CD20, partially positive for MUM1, BCL6, and negative for CD3, CD56, ALK1, CD10, CISH-EBER, KI67-was close to a 100%.

PET-CT did not demonstrate any pathology. Treatment with the R-CHOP regimen was immediately initiated, emphasizing the importance of systemic therapy, rather than a CNS-directed therapy, in this disease entity. The patient has astoundingly recovered from GCS of 7 to full consciousness withing 1 cycle. All in all, the patient=was treated with 6 cycles of R-CHOP and four intercalating cycles of high-dose methotrexate and attained Complete Remission (CR) clinically and radiographically. Due to disease severity and known poor prognosis, she was consolidated with thiotepa-based high-dose therapy and autologous stem cell transplantation. She is in CR a year after the end of therapy. Although she has regained most of her cognitive and motor functions, she does tend to be forgetful, has difficulty with naming and complex task management. She is ambulatory, but still suffers from residual peripheral neuropathy.

Discussion

Diagnosis in B-cell Intravascular is made post-mortem in about half cases [4]. Cognitive impairment, as observed in this patient,

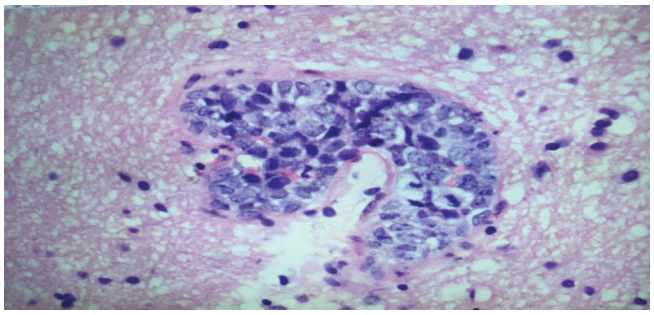

Figure 2 Expansion of vascular cavities of large cell clusters in the vascular space, compatible with intravascular large B cell lymphoma with hemorrhages (H&E stain).

occurs in 60% of patients with CNS involvement. Other symptoms are paraplegia (22%), seizures (13%), and vision disturbances (8%), although any symptoms may occur, depending on the specific site of vascular occlusion [5]. Neuroimaging findings in patients with IVL can vary from patient to patient. In a case series five MRI patterns were described: infarct like lesions, nonspecific white matter lesions, meningeal enhancement, and mass like lesions, and hyperintense lesions in the pons on T2WI [6]. None of them had bleeding on MRI.

There are only two case reports of IVL mimicking AHLE. The first was diagnosed post-mortem, and no SWI sequence was reported [7]. The second was in a patient with progressive neurological

syndrome with partial seizures and focal deficits, who died during hospitalization due to cardiopulmonary arrest. AHLE was suspected because of bleeding shown in Gradient echo in MRI [8]. To our knowledge, there is no other report of similar SWI MRI images. In contrast to the aforementioned cases, here the rapid diagnosis enabled successful treatment.

There are few clues that led to the diagnosis of IVL: The patient's age, fever, and the laboratory findings of anemia and elevated LDH are compatible to the diagnosis of IVL. On the other hand, bleeding of any kind (including AHLE) can cause elevated LDH, and the pleocytosis in the CSF in AHLE is more commonly PMN but can be MN as well. The negative total CT, and the negative skin biopsy, cannot rule out IVL but certainly lower the likelihood of the diagnosis. As to our MRI, it could not differentiate between IVL to AHLE: the lesions in AHLE are typically in the white matter, hemorrhagic and diffuse-all of which were present in our patient, but IVL can mimic almost any diagnosis. Only biopsy can differentiate between the two.

Conclusion

Our conclusion is that in order to diagnose IVL with CNS involvement in time, high level of suspicion should be maintained. Moreover, when CNS involvement is suspected, a brain biopsy should be taken as soon as possible. In our case, the quick biopsy that was preformed immediately after the MRI has changed the course and prognosis of this woman.

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