

Rapidly progressive neurological deterioration without magnetic resonance imaging abnormalities: intravascular lymphoma



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50-year-old man with a negative medical history presented with aphasia and influenza-like symptoms over the course of 2 days. He was transferred to our university hospital because of unexplained encephalopathy with mutism and apraxia. Laboratory analysis revealed an elevated lactate dehydrogenase level of 1282 U/L (reference range, <250 U/L) and a normal leukocyte count with normal differentiation. Cerebrospinal fluid examination was negative for an infection, and only a few normal T cells were present. Surprisingly, magnetic resonance imaging of the brain (panel A) did not show any abnormalities, and computed tomography also failed to show lymphadenopathy. Unfortunately, the patient developed respiratory insufficiency after a seizure and was then intubated. A bone marrow smear showed large blast-like cells with prominent nucleoli (panel B), and immunophenotyping showed a small CD45 weak population (2%) positive for CD19, CD20, and HLA-DR and negative for CD34, CD10, T markers, and myeloid markers.

Treatment with rituximab-cyclophosphamide-doxorubicin-vincristin-prednisone was directly initiated, and in less than a day, the patient was detubated. A few days later, the bone marrow biopsy demonstrated intravascular cells positive for CD20 (panels C and D) and negative for a c-myc translocation, with a high Ki-67 proliferative index (>90%). This confirmed the diagnosis of intravascular lymphoma. The patient was discharged within a week after extubation.



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