Malignant cerebral edema: an unusual neurological manifestation of systemic lupus erythematosus

Edema cerebral maligno: uma manifestação neurológica atípica do lúpus eritematoso sistêmico

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A 46-year-old woman presented with headache and decreased level of consciousness. She had systemic lupus erythematosus (SLE) with positive antibodies. Cerebrospinal fluid (CSF) pressure was 350 mm ${\rm H_2O}$. Brain magnetic resonance imaging (MRI) revealed white matter changes and edema. MRI angiography and vessel wall imaging ruled out vasculitis (Figure 1). Malignant cerebral edema related to SLE was diagnosed. She had complete recovery after methylprednisolone and cyclophosphamide.

Several neurological manifestations have been described in SLE, including intracranial hypertension syndrome, which is unusual¹. Malignant cerebral edema is a distinct syndrome rarely observed in SLE². Severe bloodbrain barrier disruption and vasculitis are the most likely pathophysiological mechanisms, and treatment includes immunotherapy².

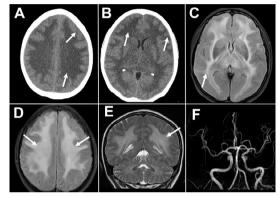


Figure 1. Brain CT scan shows cerebral edema with loss of sulci and white matter hypodensity (A and B). Axial FLAIR-weighted and coronal T2-weighted brain MRI reveal bilateral, symmetrical, and diffuse hyperintense signal in white matter, with loss of cerebral sulci, characterizing cerebral edema (C, D, and E). MRI angiography is normal (F).

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Conflict of interest: There is no conflict of interest to declare

Authors' contributions: MMPM: case report project conception, organization, execution, manuscript writing of the first draft, review and critique. BT: case report project: conception, organization, execution, manuscript writing of the first draft, review and critique. FMRF: case report project: conception, organization, execution, manuscript review and critique. VHRM: case report project conception, organization, execution, manuscript review and critique. OGPB: case report project conception, organization, execution, manuscript review and critique. JLP: case report project conception, organization, execution, manuscript review and critique.

Received on May 26, 2020; Received in its final form on June 5, 2020; Accepted on June 15, 2020.



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