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Encephalitis with Perivascular Enhancement

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Clinical image description

Hodgkin lymphoma represents 10% of all lymphomas, with nodular sclerosis subtype being the most common. Direct involvement of the central nervous system as a complication of Hodgkin lymphoma is rare, it has been described in 0.5% of patients, indirect involvement can manifest itself as paraneoplastic, infectious or inflammatory [1].

We present a 46-year-old caucasian man with a history of hypertension and stage IV-B nodular sclerosis Hodgkin lymphoma (with lung and bone involvement) in complete remission after treatment with 6 cycles of chemotherapy.

The patient presented a generalized tonic-clonic seizure lasting 40 minutes that did not respond to midazolam and required intubation due to low level of consciousness. Treatment with levetiracetam and valproate infusion was started. During the previous week he had presented headache, dizziness and some vomiting without fever or other symptoms. An electroencephalogram was performed showing no focal or diffuse paroxysms suggestive of epileptic etiology. A brain Magnetic Resonance Imaging (MRI) showed confluent bilateral and left temporal fronto-parietal white matter lesions and multiple perivascular linear enhancements in practically the entire white matter in post-contrast sequences, and angiographic study showed no stenosis (Figure 1). A lumbar puncture was performed, evidencing hyperproteinorrhea and lymphocytosis with a predominance of polymorphonuclear cells without atypia and a negative microbiology.

The patient was oriented as a vasculitis of probable paraneoplastic vs infectious etiology. A spectacular clinical and radiological response to corticosteroid treatment was observed, orienting the case towards paraneoplastic vasculitis. The brain MRI showed disappearance of the majority of previous brain lesions and disappearance of lesion enhancement one month later (Figure 2). Anti-GFAP (Glial Fibrillary Acidic Protein) antibodies were not obtained due to unavailability. Seven years later the patient remains stable with well-controlled lymphoma and in remission.

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In this clinical case, neuroimaging with post-contrast perivascular enhancement and response to corticosteroids were key in the differential diagnosis that includes infectious (Herpes virus, listeria), paraneoplastic (anti-GFAP encephalitis, paraneoplastic cerebellar degeneration), tumoral (primary or secondary nervous system lymphoma) and inflammatory etiology (granulomatous angiitis of the central nervous system, CLIPPERS - Chronic Lymphocytic Inflammation with Pontine Perivascular Enhancement Responsive to Steroids) [2].

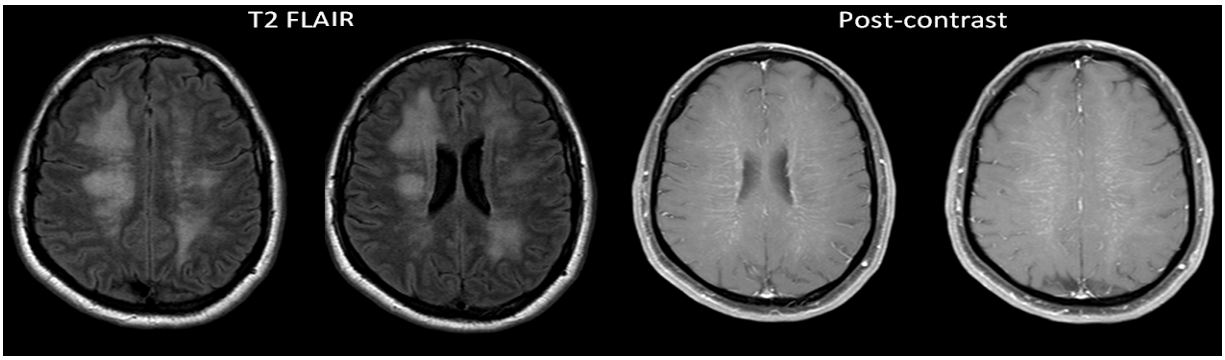

Figure 1: Brain MRI.

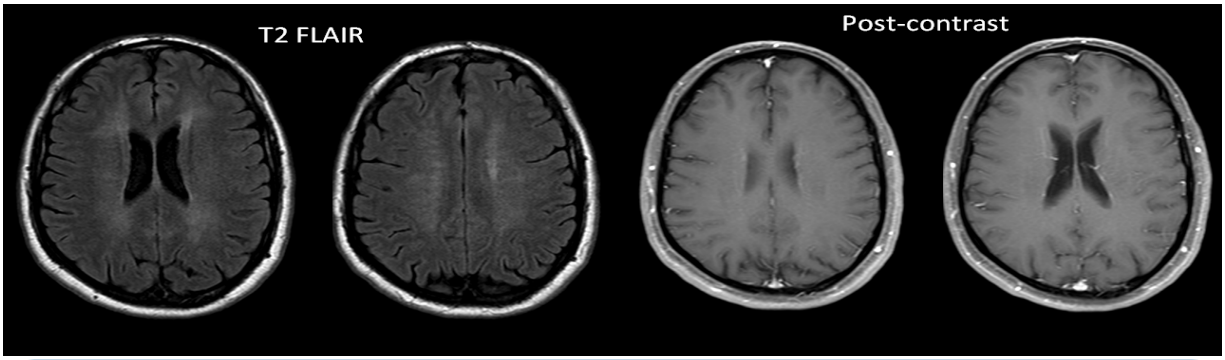

Figure 2: Brain MRI.

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