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Pseudotumoral presentation of primary central nervous system vasculitis

Vasculite do sistema nervoso central na forma pseudotumoral

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CASE REPORT

A 28-year-old man, with insidious and progressive onset of semantic and phonemic aphasia, associated with evolving numbness and loss of sensation over weeks. Magnetic resonance imaging (MRI) revealed T2-hyperintense expansive lesions, heterogeneous contrast enhancement, and no diffusion restriction in subcortical regions on the left parietal and temporal lobes, right superior parietal lobe, post-central gyrus and splenium of *corpus callosum*, with perilesional edema and hemorrhagic foci. Steroid and cyclophosphamide pulses were introduced and stabilized the clinical symptoms.

One month later, a new MRI (Fig 1) showed improvement of the expansive effect, edema and enhancement of the lesions. Magnetic resonance angiography (MRA) and digital subtraction angiography (DSA) presented only a subtle enhancement in the capillary-venous phase in the left frontal and parietal lobes.

The brain biopsy was performed and revealed post-ischemic necrosis associated with nonspecific perivascular lymphomonocytic infiltrate with predominance of T lymphocytes without atypias. Immunohistochemistry showed small numbers of mature perivascular lymphocytes, CD20+, CD3+ and CD68+ cells, which are compatible with lymphocytic vasculitis. Clinical and pathological picture was compatible with primary lymphocytic angiitis of the central nervous system (PLACNS).

Corticosteroid (1 g/day) and cyclophosphamide (1 g/day) pulse therapy was introduced for three days and repeated monthly during three months. Prednisone (20 mg/day) was introduced as maintenance therapy, and resulting in improved clinical outcomes and MRI imaging. In the fifth month, the patient presented worsening of the sensitive symptoms and developed new MRI lesions (Fig 2).

New corticosteroid and cyclophosphamide pulse therapy was introduced for three months, resulting in almost complete resolution of the symptoms and improvement of MRI findings. Prednisone 40 mg/day was introduced, but no improvement in the MRI was observed. Later, azatioprine (50 mg/day) was associated with the therapy. A one-year follow-up MRI showed significant improvement of the lesions (Fig 3).

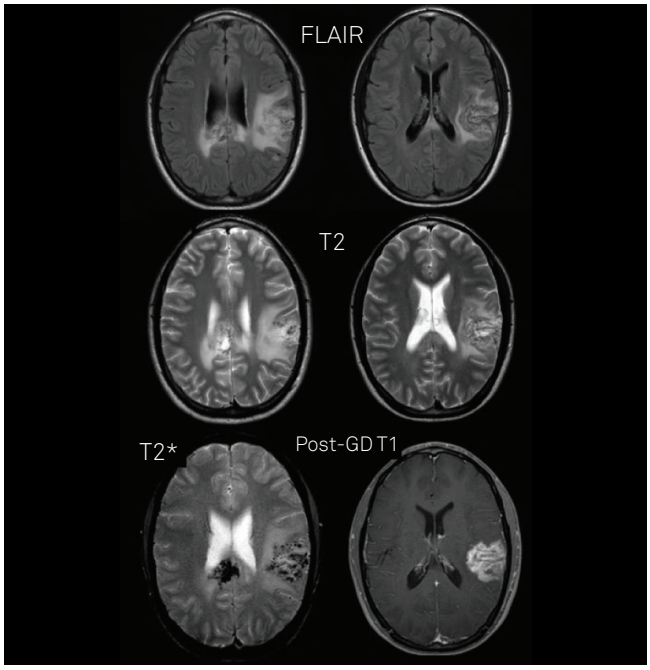

Fig 1. Brain magnetic resonance imaging one month after the onset of clinical symptoms and corticosteroid therapy. Expansive lesions were detected in left cortical/subcortical parietal lobe and in the splenium of the *corpus callosum*. The lesions presented heterogeneous hyperintense signal on FLAIR and T2-weighted images, marked hypointense signal on T2* (blood products), and contrast enhancement (blood brain barrier disruption). Perilesional edema was also noted. The other brain lesions detected presented the same signal intensity (not shown).

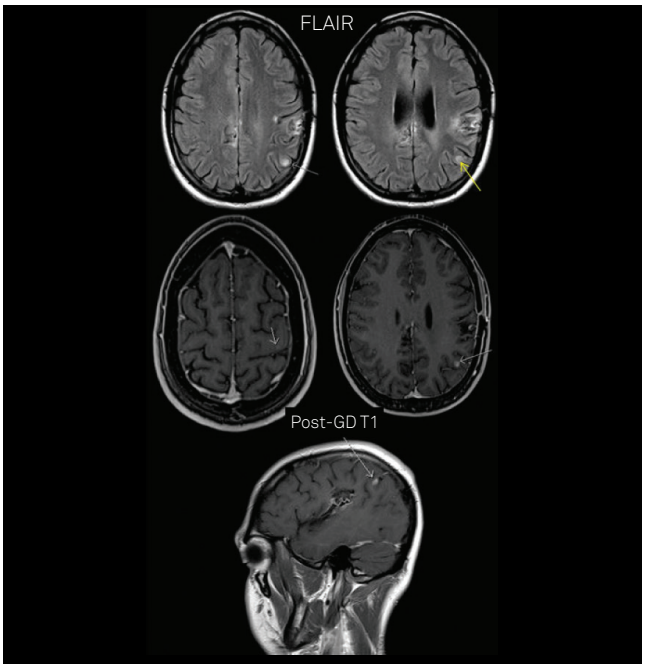

Fig 2. Five months after the onset of symptoms (one month after corticosteroid-cyclophosphamide pulse therapy), the patient presented clinical worsening. New enhancement foci were detected in the left pre and post-central gyri. The initial parietal lesion present on this examination decreased in size, and the enhancement and perilesional edema decreased when compared to the prior examination (Fig 1).

DISCUSSION

PLACNS is a rare inflammatory vasculitis that affects the parenchyma and meningeal arteries and veins[1-4]. An even less common subtype of PLACNS is the lymphocytic vasculitis[1,5]. Pseudotumoral presentation corresponds to 5% of PLANCS. The pathogenesis, diagnosis, and treatment of the disease are challenging with few reports in the literature[3,5]. Its cause remains unclear, but a viral etiology, as well as an association with lymphoma, has been suggested[1,3]. If not treated, progressive neurological decline or even death may occur[1,4].

PLANCS is usually diagnosed by exclusion based on radiological, pathological, and immunohistochemical findings[5-7]. Three main histological patterns have been reported, the granulomatous inflammatory pattern, the lymphocytic pattern, and the acute necrotizing one[3,8].

MRI, MRA, and DSA can help in the diagnosis[1,4]. Proton spectroscopy can help excluding other pathologies in case of pseudotumoral form[1]. MRI findings are almost always present, but they are inespecific[2]. Diffuse white matter lesions and microangiopathy are commonly observed[2,6,9]. Despite a positive biopsy, DSA can be normal if small brain vessels are affected[4,9]. On the other hand, a negative biopsy cannot exclude vasculitis[4]. When clinical and MRI findings

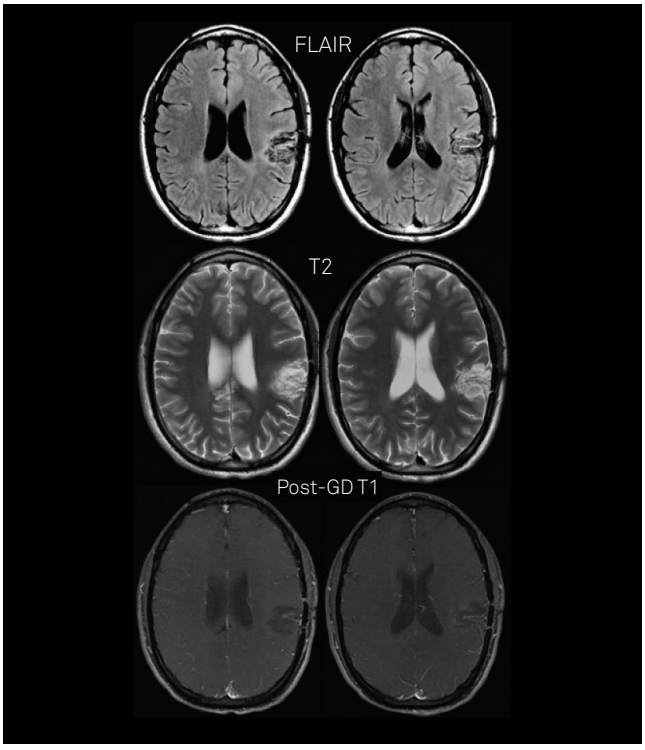

Fig 3. One-year follow-up magnetic resonance imaging. The patient was asymptomatic. Sequelar lesions were detected, without enhancement or mass effect.

suggest the diagnosis of PLACNS, an immunosuppressive therapy is indicated[4,10,11].

The studied patient presented the pseudotumoral form of PLACNS with MRI abnormalities, negative DSA, and positive biopsy. One-year-follow-up showed complete resolution of symptoms and MRI improvement, without contrast enhancement.

PLACNS diagnosis is challenging, and it requires clinical history, laboratorial findings, imaging, and eventually a histopathological confirmation.

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