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Challenging presentation of primary vasculitis of the central nervous system

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A B S T R A C T

Objective: To show a patient with a diagnosis of primary vasculitis of the nervous system and review this entity's literature.
Clinical case: Male 32 years, with two events; first, with a transient monoparesis of the right upper extremity, improve with steroids in days. Now, with a motor Jacksonian progression from the upper to the lower right extremities and generalized seizures. After; aphasia, right hemiparesis, and delirium. In the antiresonance, the finding of multiple arterial cerebral "beading." With an in-depth study, the diagnosis of primary cerebral vasculitis was made. The management with levetiracetam, steroids, and Azathioprine offers a satisfactory evolution.
Discussion: The primary CNS vasculitis is an exclusion diagnosis, with angio-MRI is possible to suspect it, but it is always obligate to discard a secondary etiology by infection, systemic diseases, neoplasia, and drugs.
Conclusion: Although this problem is infrequent, we must consider this possibility. Opportune treatment can restore the quality of life.

Background

Primary cerebral vasculitis refers to an inflammation within the wall of the central nervous system (CNS) blood vessels associated with destructive changes rather than occlusion and infarction [1,2]. Unlike Secondary CNS vasculitis, which usually results from infections, connective tissue diseases, neoplasms, and drugs, primary cerebral vasculitis is considered to occur with no identified etiology [1–3] CNS damage was reported in 24% of primary systemic vasculitis cases, commonly due to the antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis and polyarteritis nodosa [2]. Conversely, in "primary" or "isolated" vasculitis or angiitis of the CNS, there is little or no overt generalized inflammation [2–4]. The range of clinical expressions of primary CNS vasculitis damage is relatively wide. The clinical presentation related to the site of the vasculature affected, including headache, cognitive change, and generalized seizures, the focal neurological abnormalities are also common, including hemispheric, brainstem or spinal deficits, movement disorders, and optic and other cranial neuropathies secondary to cerebrovascular damage such as hemorrhagic and ischemic stroke caused by intra/extra-cerebral vascular stenosis, aneurysm, and sinus venous thrombosis, meningeal and brain parenchymal involvement resulted from granulomatosis and perivasculitis, and encephalopathy due to cytokine damage [1–7]. As with the clinical features, so for investigations: there are no biochemical, immunological or serological, or imaging investigations that are diagnostic of primary CNS vasculitis [1,2,8,9]. The proposed criteria for the diagnosis of central nervous system vasculitis: Definite (a) Clinical presentation suggesting CNS vasculitis with the exclusion of possible alternative diagnoses and primary systemic vasculitic syndrome (b) Plus the presence of characteristic CNS histology, that is, biopsy or autopsy showing CNS angiitis, including evidence of vessel wall damage. Possible diagnosis characteristics are a) Clinical presentation suggesting CNS vasculitis with the exclusion of possible alternative diagnoses and primary systemic vasculitic syndrome b) Plus laboratory and imaging support for CNS inflammation (elevated levels of cerebrospinal fluid protein and cells, and the presence of oligoclonal bands and MR (Magnetic resonance) scan evidence compatible with CNS vasculitis), with the angiographic exclusion of other specific entities, (c) But without histological proof of vasculitis [1,2,10]. To date, the treatment of primary CNS vasculitis does not have an evidence base from direct clinical trials. However, there is mention in the literature that immunosuppressants continue to be the cornerstone of treatment, such as steroids, cyclophosphamide, Azathioprine, and methotrexate are

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administered for weeks as an induction period. The use of monoclonal antibodies such as rituximab has been mentioned; however, more studies are necessary to confirm the efficacy of this drug [1–3,11,12].

Case presentation

We are reporting a 32-year-old Mexican male without significant antecedents. In 2018 the patient presented an episode of monoparesis of the right upper limb. Without identification of the cause, the patient showed partial improvement in the force. In October 2020, the patient developed bitemporal headache of moderate-intensity, intermittent, and with improvement associated with the use of analgesics as well as right faciobrachial paresis together with numbness in the same location, adding to the following hours' paresis of the right lower limb as well as dysarthria as well as generalized tonic-clonic seizures on three occasions, which is why the patient goes for medical evaluation. The neurological examination showed the patient inattentive, evaluating the cranial nerves, the right supranuclear facial palsy, strength 2/5, hyperreflexia, and extensor plantar response in the right hemibody, as well as hypoesthesia in the exact location, were highlighted.

Investigations

Routine laboratory studies were performed where glycemia 403 mg/dl, calcium 11 meq / l, magnesium in 5 meq / l, pH 7.3, HCO3 15, Lactate 15 stood out, values that improved with subsequent hydration. The following studies, Human immunodeficiency virus, hepatitis *C* virus, hepatitis *B* virus, cell erythrocyte sedimentation rate, C-reactive protein, thyroid-stimulating hormone, t4, antinuclear antibodies, and ANCAS, complete and rheumatoid factor, were negative. The cerebrospinal fluid only showed proteins of 81.7 mg/dl. Gadolinium MRI was performed. Hypointense and hyperintense lesions were observed in T1 and T2, respectively, and the restriction in ADC (Apparent diffusion coefficient) and diffusion of lesions in the centrum semiovale parietal region in a bilateral manner, without contrast uptake of these lesions. (Fig. 1)

Besides, he presented a bulging in the general distribution of the arteries and a narrowing of the distribution of the middle cerebral artery bilaterally, seen both on CT angiography and MR angiography. (Fig.2)

Differential diagnosis

Various studies were performed, such as blood tests, CT images of the chest, abdomen, pelvis, CSF examination, and MRI, to rule out other alternative disorders such as autoimmune disorders, infectious and malignancy; however, the presence of any other disease was not evidenced. In addition to the use of MRI, it was considered to perform non-constant digital subtraction angiography since this last study has been associated with a significant risk of developing stroke, so it was decided not to use this study. We rely on the criteria of possible disease to avoid taking a biopsy.

Treatment

In the current literature, certain immunosuppressive drugs such as steroids, cyclophosphamide, Azathioprine, and methotrexate are determined as the main drugs used in primary vasculitis of the central nervous system. In the case of our patient, steroids and Azathioprine were used.

Outcome and follow-up

Our patient showed an adequate response two weeks after being hospitalized using the steroid and Azathioprine.

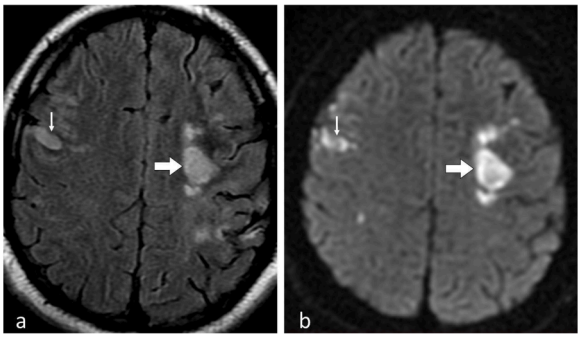

Fig. 1. Axial FLAIR T2 (a) and DWI (b) show subacute-chronic, cortical and subcortical cerebral stroke in both frontal lobes that restrict diffusion.

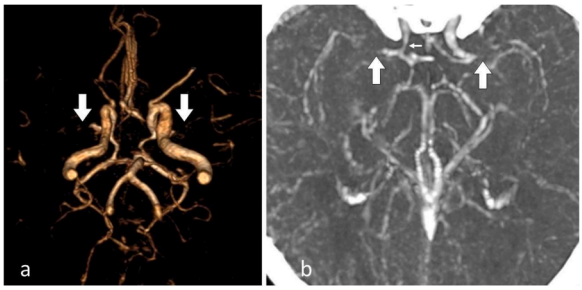

Fig. 2. 3D TOF MRA (a) and CT angiography (b) shows irregularities and occlusion of the middle cerebral arteries bilaterally, predominantly in their proximal M1 segments. The right internal carotid artery is smaller in diameter compared to its contralateral counterpart. Findings consistent with vasculitis.

Discussion

Primary central nervous system vasculitis (PCNSV) is an infrequent inflammation of the arterial branches. Two possible mechanisms could be involved: the deposition of immune complexes and cell-mediated autoimmunity, resulting in immune inflammation and necrosis of the wall from the vessels. This histopathological finding affects people of any age but is more frequent between the fourth and sixth decade of life; Brain and meningeal biopsy is the gold standard for diagnosis, with sensitivity ranging from 50–70%. A biopsy of the brain may be falsely negative when the not-affected tissue is sampled; it is a procedure that carries risk, and it is not easily be repeated over time. Bleeding at the biopsy site is the most frequent complication (4.9 %); however, permanent neurological sequelae also occur at a lower frequency (1 %) [13]. In the histopathological diagnosis, the presence of transmural inflammation with injury to the vessel wall is required; usually, there is an angiocentric inflammatory infiltrate. However, the improvement in image quality has been superseded, including subtraction angiography, and may not require biopsy if sufficient clinical evidence is available [1–3].

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