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Primary central nervous system vasculitis disguised as tumor-like granulomatous angiitis and multifocal subdural hematomas: A case report and literature review

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ABSTRACT

Primary central nervous system vasculitis (PCNSV) is a rare inflammatory disorder limited to the brain and/or spinal cord that destroys CNS vessels without evidence of vasculitis outside the CNS. We present a case of a 52-year-old male who complained of a tension headache and dizziness. He was diagnosed postbiopsy with PCNSV that was initially disguised by a tumor-like lesion and multifocal subdural hematomas seen on MRI. Our report highlights the value of considering PCNSV as a differential diagnosis in patients with subdural hemorrhage that has never been previously recognized and emphasizes the value of a brain biopsy for confirming the diagnosis. Our report also shows the efficacy of steroid and immunosuppressive therapy in treating PCNSV.

1. Introduction

Primary central nervous system vasculitis (PCNSV) is a rare disorder that is usually considered a diagnostic challenge. The average age of onset of this condition is approximately 50 years, but it may affect patients of all ages. Neurological manifestations are various, the most common being a headache, cognitive deterioration, and focal neurological deficits such as intracranial hemorrhage. Unfortunately, the cause and pathogenesis of PCNSV are unknown [1].

In PCNSV, the inflammatory process is confined to the CNS only. No single lab test has a satisfactory sensitivity or specificity for establishing the diagnosis, and high clinical suspicion is necessary. Obtaining a brain biopsy is required to confirm the diagnosis and exclude other causes. PCNSV is a notoriously patchy disorder, which limits the sensitivity of the procedure, and as many as 25% of the obtained biopsies are falsely negative. The presence of angiitis in the biopsy specimen should not prevent the performance special immunohistochemistry stains and cultures for underlying infections that can elicit secondary vascular inflammation [1].

We present a unique case of PCNSV manifesting as a steroid responsive tumor-like granuloma associated with subdural hemorrhage with distinctive findings that have never been previously reported. This case also emphasizes the value of a brain biopsy for confirming the diagnosis and highlights the efficacy of steroid and immune suppressive therapy for treating PCNSV.

2. Case study

A 52-year-old male with known juvenile diabetes mellitus presented to our emergency room complaining of dizziness and a progressive tension headache that had not responded to analgesics for one week. There was no history of fever, convulsions, photophobia, vomiting or weight loss. He denied any history of trauma. He was vitally stable, fully alert and oriented. The remainder of his clinical exam was within normal limits. A laboratory workup showed no abnormal findings. Similarly, his serum chemistry results were within the normal ranges.

A diagnostic CT scan revealed the presence of a well-defined hyperattenuating lesion in the left temporal lobe and bilateral extra-axial hyperattenuation overlying the tentorium cerebelli with suspicion of hemorrhage (Fig. 1). Furthermore, an MRI revealed an intra-axial bubbly lesion in the left temporal lobe that showed high T2/FLAIR and T1 signal intensity with no enhancement after a gadolinium injection. Moreover, multiple bilateral extra-axial lesions with high FLAIR and T1 signal intensity and low T2 signal intensity consistent with multifocal subdural hematomas (Fig. 2). Nevertheless, an exploratory spinal MRI was unremarkable. These characteristics were in favor of extra-axial dural hemorrhagic metastasis. Another differentials were metastatic melanoma and infectious encephalitis. A diagnostic lumbar puncture

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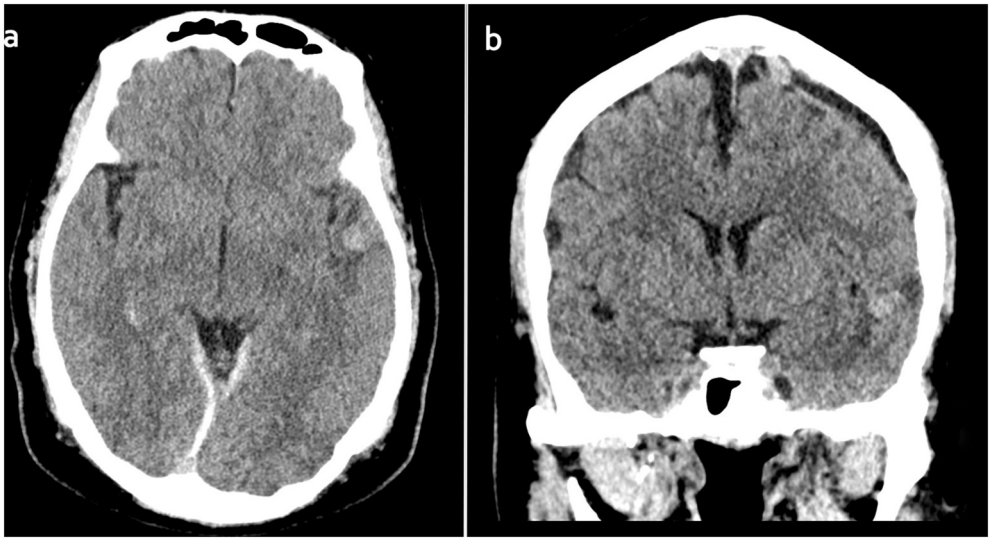

Fig. 1. Transaxial [a] and coronal [b] unenhanced CT scan of the brain, exhibits a well-defined intra-axial hyperattenuated lesion in the left temporal lobe with mild perilesional edema. There is bilateral subdural hyperattenuation overlying the tentorium cerebelli, which represents a subdural hemorrhage [a].

was performed and showed standard CSF parameters; moreover, blood tests for antineutrophil cytoplasmic antibody (ANCA) and antinuclear antibody (ANA) were negative.

Subsequently, the patient underwent a left temporal craniectomy and excisional biopsy that revealed a consensus findings of PCNSV based on immunohistochemical analysis (Fig. 3). By combining these findings with the radio-clinical assessment, the diagnosis of vasculitis was favorable. As a consequence, the patient was treated with steroids and immunosuppressive therapy and showed significant improvement in his symptoms (Fig. 4). Later, the patient was discharged from the hospital in good health.

3. Discussion

PCNSV is a granulomatous inflammatory disorder that is often disguised by other clinical conditions, such as a migraine, stroke, epilepsy, dementia, demyelinating disorders and CNS infections, which make PCNSV a diagnostic challenge [1]. Although nonspecific, all cases have abnormal MRI findings that suggest the possibility that PCNSV cannot be excluded, even with normal angiography. Although noninvasive, abnormal magnetic resonance angiography is less sensitive for detecting vasculitic arterial changes, which may be present in other systemic forms of vasculitis with CNS involvement and other nonvasculitic conditions [2].

There is no consensus on diagnostic criteria, and most clinicians still use the criteria that Calabrese and Mallek [3] proposed over 20 years ago linking the clinical diagnostic workup with imaging/histopathologic abnormalities and excluding other clinical attributes. Our patient fulfilled all three measures for diagnosing PCNSV. He presented with a severe headache, which is the most common symptom encountered in PCNSV patients [4]. Our patient's headache was tension in nature, with increasing intensity, and did not respond to analgesic medications.

An MRI revealed multiple hemorrhagic lesions. It is essential to recognize that intracranial bleeding involving intracerebral and subarachnoid hemorrhage can occur in approximately 12% of PCNSV patients because the early recognition and treatment of the underlying vasculitic process may help avoid severe outcomes [5]. Our case appears to be the first reported case of PCNSV with subdural intracranial hemorrhage. Furthermore, on an MRA our patient showed bilateral narrowing of both the anterior and middle cerebral arteries with a beaded appearance of the side wall configurations (Fig. 2).

Invasive biopsy of the brain and/or spinal cord is considered the gold standard to confirm the diagnosis of PCNSV. In summary, it is important to recall that biopsy is used not only to provide pathological proof of the diagnosis of vasculitis but also to exclude alternative diagnoses. The immunohistochemical analysis of our lesion revealed unique features, since the most common histopathological pattern of PCNSV associated with intracranial hemorrhage is acute necrotizing vasculitis [2].

Histopathological analysis revealed the presence of an immune-mediated reactive process, as evidenced by foci of inflammatory cells and lymphocytic, eosinophilic and neutrophilic infiltrations, mixed with an element of gliosis and foamy macrophages with a faintly vacuolated cytoplasm. These findings in small vessel walls and in the perivascular region are characteristic of vasculitis (Fig. 3). In particular, supportive clinical and imaging findings and the detection of lymphocytic infiltration in and/or around the walls of parenchymal or leptomeningeal vessels are the primary findings in CNS vasculitis [6].

One differential diagnosis for this case was infectious encephalitis, such as herpes simplex encephalitis (HSE); immune-mediated demyelinating diseases might also mimic this condition. Nevertheless, as the patient was a middle-aged male with an acute onset of symptoms, no history of fever, altered sensory neuron deficits, focal neurological deficits or seizures, and no abnormal CSF parameters, HSE was unlikely [7]. Moreover, the unsupportive imaging findings and signal characteristics of the multifocal axial and extra-axial lesions on MRI (Fig. 2) excluded the possibility of a demyelinating disease. In addition, the finding of a narrow and beaded appearance on MRA supported a diagnosis of vasculitis.

Differentiating these entities based on histopathology alone is quite difficult, as HSE commonly shows massive tissue necrosis with hemorrhage [8], which can also be identified in 25% of patients with necrotic vasculitis [4]. Nevertheless, the combination of these findings with the provided diagnostic criteria [3] made the diagnosis of vasculitis favorable. Moreover, the dramatic regression of the patient's clinical condition and imaging findings without any treatment with antiviral agents but only with steroids and immunosuppressive therapy excluded HSE (Fig. 4).

Our patient showed significant improvement after establishing the diagnosis and starting a regimen of aggressive immunosuppressive treatment. Recognizing the possibility of PCNSV may avoid the need for excision of mass lesions. Because of potential PCNSV mimics, revisiting