

Massive Bitemporal Hemorrhages in Antineutrophil Cytoplasmic Autoantibodies Vasculitis

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ANCA-associated small vessel vasculitides are systemic diseases characterized by inflammation and necrosis of small-sized blood vessels.¹ They are characterized by the presence of antineutrophil cytoplasmic antibodies (ANCA), which primarily target the kidney, lung, skin and peripheral nervous system.² In rare cases, they can affect the central nervous system (CNS) and cause cerebrovascular events.³ We present a patient with ANCA vasculitis who developed fatal massive bitemporal hemorrhages during the course of immunosuppressive therapy.

A 71-year-old man was referred to our tertiary neurological center due to weight loss (10 kg in two months), fatigue, weakness and persistent fever of unknown origin. The patient did not report previous sinusitis, upper airway infection, arthritis or hematuria. Neurological examination only revealed a right foot drop. Laboratory studies showed inflammatory anemia and elevated inflammatory markers. Screening for infectious diseases was negative. Spine MRI indicated a herniated L5-S1 disc. Brain MRI was normal except for a millimetric left occipital ischemic infarct. Whole-body PET scan was conducted to eliminate a neoplastic etiology, but was also negative. Five days after his arrival, the patient developed new weaknesses and paresthesia in the left leg and right hand. Laboratory studies revealed acute renal failure (creatinine 133 $\mu\text{mol/L}$, hematuria, proteinuria) and elevated c-ANCA counts (1:320). A second MRI of the brain conducted 10 days after the first showed five new millimetric ischemic foci in the right temporal and left cerebellar regions. In concert with rheumatology, a diagnosis of ANCA vasculitis was made, an immunosuppressive treatment was initiated (high-dose intravenous glucocorticoids), and the patient's neurological symptoms were stabilized. However, creatinine continued to rise (from 133 to 177 $\mu\text{mol/L}$ in 7 days). A renal biopsy was scheduled to confirm a diagnosis of glomerulonephritis. After five days of induction treatment, tapering oral doses of prednisone were administered. On the first day of prednisone tapering, the patient was found unresponsive in his bed (eyes opened, but unresponsive to simple commands). Head CT showed massive bilateral temporal hemorrhages (see Figure 1). After discussion with the family, the patient was transferred back to his hometown for end-of-life care. Renal biopsy and autopsy could not be performed. Nevertheless, a recent meta-analysis has evaluated the specificity of elevated c-ANCA levels in the serum at 96.3% (CI [94.1-98.5]),⁴ which, combined with the high pretest probability of vasculitis (mononeuritis multiplex, acute renal failure, inflammatory anemia,



Figure 1: Axial view of a head CT showing massive bitemporal haemorrhages with significant mass effect.

multifocal brain infarcts), made a diagnosis of ANCA vasculitis highly probable even without a gold-standard pathological diagnosis.

While it is known that ANCA vasculitis may in some instances involve the CNS,^{3,4} only a few case reports describe the appearance of cerebral hemorrhages,⁵⁻¹⁰ and even fewer a rapid progression toward massive hemorrhages and death.⁵⁻⁷ Altogether, our case supports the possibility of cerebral

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vasculitis, though rare in ANCA-positive patients, causing so massive a cerebral hemorrhage.

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STATEMENT OF AUTHORSHIP

David Bergeron drafted the initial version of the manuscript. Dr. Annie Dionne was the treating physician. All authors contributed equally to the writing process and approved the final version of the manuscript.

DISCLOSURES

David Bergeron, Raphaël Marchand and Robert Jr Laforce do not have anything to disclose.

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