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More than meets the eye. Antineutrophil cytoplasmic autoantibody (ANCA) – associated vasculitides (AAV) – a diagnostic challenge in elderly

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Case Study

Introduction: Patients with AAV may present with a variety of signs and symptoms, and renal and pulmonary involvement is common. However, the small vessels in any organ or tissue can be affected, which sometimes can mimic well-defined pathologies, causing an initial misinterpretation. Case 1: A 72-year-old male, with history of hypertension and dyslipidemia, was admitted into the Stroke Unit after presenting with a motor deficit of the right upper limb, with onset 3 hours before. Brain CT was normal. Laboratory assessments showed AKI KDIGO 1 and eosinophilia ($0.9x10^9$/L). At day 2, the patient developed motor deficit of the left arm and progressive AKI. Subsequent tests showed: active urinary sediment, non-nephrotic proteinuria, MPO-ANCA titles 119UI/mL. The diagnosis of MPO-AAV was made and confirmed with kidney biopsy. The patient received steroid pulses followed by oral prednisone and rituximab. Kidney function and neurologic symptoms improved and, 1 year later, he is in remission.

Case 2: A 81-year-old male, with history of CKD 3a, DM and hypertension, presented to the ER with precordial pain and palpitations. He also reported anorexia, nausea and vomiting during previous days. Electrocardiogram showed no ST-elevation. Laboratory assessments showed anemia, cardiac troponin I 36400ng/L and AKI KDIGO 3. He was admitted to the Coronary Intensive Care Unit with an acute myocardial infarction. There was no improvement of AKI after hemodynamic stabilization and subsequent analysis showed: active urinary sediment, nephrotic proteinuria and MPO-ANCA titles 127UI/mL. Diagnosis of MPO-AAV with kidney and heart involvement was made. Treatment included steroid pulses followed by oral prednisone and intravenous cyclophosphamide. Kidney biopsy was not performed due to CKD. Unfavorable kidney evolution occurred, leading to hemodialysis requirement.
Conclusion: These cases highlight the importance of valuing uncommon major organ involvement as main manifestation in AAV. Given the rarity of the pathology, a high clinical suspicion is mandatory.