VASCULAR DISEASE, VASCULITIS

AN UNUSUAL PRESENTATION OF GRANULOMATOSIS WITH POLYANGIITIS

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Background: Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a granulomatous necrotizing vasculitis that is part of small vessel vasculitis associated with the presence of ANCA antibodies. It is a rare pathology of unknown etiology involving in its classical form upper and lower airways and kidney.

Observation: We report a case of a 19-year-old woman who was admitted to our hospital, with a 6-month history of anemia, asthenia, anorexia, adinamia and weight loss, associated with multiple episodes of epistaxis and proctorrhagia. Physical exam revealed erythematous and painful papules and plaques, and hemorrhagic blisters, acompanied by focal necrosis and ulceration in the lower extremities. Perimalleolar edema and arterial hypertension were present. Blood test showed Hto 28%, Hb 8 gr/dl, MCV 64, ESD 55 mm/hs, CRP 41 mg/L, ALP 420 IU/L and ANCA -C positive. Urinalysis was normal. Computed tomography scans revealed mucosal thickening of frontal, sphenoid and maxillary sinuses, pulmonary nodules and bilateral renal enlargement with hypodense foci in the renal parenchyma that do not enhancement with intravenous contrast. A skin lesion biopsy was performed and revealed a leukocytoclastic vasculitis. Direct immunofluorescence study showed deposits of IgM and C3 in the wall of dermal vessels. CT guided percutaneous renal biopsy showed necrotizing granulomatous interstitial nephritis, confirming the diagnosis of GPA.

Key message: The purpose is to show a rare pathology that was atypically manifested, being the renal inflammatory pseudotumor an exceptional manifestation of GPA, with approximately 16 cases reported in the literature.