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ERDHEIM-CHESTER DISEASE: ACUTE KIDNEY FAILURE AS A RARE PRESENTATION OF A CATASTROPHIC DISEASE

Rita Valério Alves (1); Ivan Luz (1); Rita Calixto (3); Paulo Santos (1); Patrícia Barreto (1); Mário Góis (2); Helena Viana (2); Ana Vila Lobos (1);

(1) - Centro Hospitalar do Médio Tejo, EPE, Nefrologia, Torres Novas, Portugal;
(2) - Centro Hospitalar Lisboa Central, Nefrologia, Lisboa, Portugal;
(3) - Unidade Local de Saúde do Baixo Alentejo, Medicina Interna, Beja, Desconhecido;

A 68-year-old caucasian female was admitted to the emergency unit for dyspnea and confusional state. Past medical history included arterial hypertension, diabetes mellitus type 2, obesity, dyslipidemia, hypothyroidism, obstructive sleep apnea, pulmonar hypertension with fibrosis, a lymphoma 22 years ago with an orbital mass and loss of visual acuity that was treated only with chorticosteroids but without clinic follow-up since.
At admission, laboratory data revealed hemoglobin level 7.8 g/dL, leukocytes 23.900x109/L with neutrophils predominance, reactive C protein 7.28 mg/dL, urea 121 mg/dL, creatinine 1.7mg/dL and albumin 1.8 g/dL.
Other relevant data include: normal serum protein electrophoresis and normal serum immunoglobulins; antinuclear antibodies, antineutrophil cytoplasmic antibodies (ANCA) and anti-dsDNA antibodies were all negative. Computerized tomography (CT) of the thorax showed diffuse pleural thickening with right pleural effusion, intralobular and supleural thickening, bronchiectasis, interstitial fibrosis and circumferential sheathing of the thoracic aorta. Kidney CT showed that both kidneys were enlarged, suggesting an inflammatory process, as well as poorly delimited hypodense regions , but without hydronephrosis.
The patient started empiric antibiotic therapy with Piperaciline/Tazobactam 375mg every 6-hours, however after 10 days she evolved to a global respiratory failure, anasarca and a worsening of renal function with anuria and atrial fibrillation. The patient was transferred to the Intensive Care Unit (ICU), where she was treated with noninvasive ventilation and diuretic perfusion, to which she had no response, leading to the start of renal replacement therapy.
After reaching hemodynamic stabilization she was transferred to the Nephrology Department. She also presented an ischemia of the left inferior member (grade IV) with necessity to amputate and continued hemodialysis dependence.
Because until 1-month before the admission the renal function was normal, a renal biopsy was performed that revealed a %u201Chistiocytes population, positive for CD68 and FXIIIa, negative for S100 and CD1a%u201D suggesting Erdeim-Chester disease (ECD).
Unfortunately, the patient died before the staging of the disease was made and any therapy could be offered.
This case report is exceptional because in the existing literature all the end-stage renal disease cases report related with Erdheim-Chester disease had obstructive renal disease and ureterohydronephrosis.