Case report

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Vasculitis in elderly. About two clinical cases and literature review

Vasculitis en adultos mayores. A propósito de dos casos y revisión de la literatura

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Resumen

Actualmente vivimos un proceso de transición en la pirámide poblacional. Por lo tanto, se hacen más frecuentes las enfermedades renales en los ancianos. Así, las vasculitis primarias en adultos mayores son enfermedades raras, causadas por la inflamación de los vasos sanguíneos y muy poco diagnosticadas. La literatura se limita a escasos casos clínicos. La vasculitis por anticuerpo citoplasmático anti neutrófilo (ANCA) positivo (VAA) es más frecuente en personas mayores de 50 años. La detección de ANCA es de peor pronóstico en esta población. También, es importante la alta sospecha y diagnóstico oportuno, ya que así se puede brindar un tratamiento óptimo y disminuir las complicaciones propias de la enfermedad y aquellas asociadas a la inmunosupresión. Los adultos mayores con VAA tienen mayor compromiso renal, hipertensión, dislipidemia y mortalidad incrementada. Por lo que a continuación se detallan dos casos clínicos desde la fecha del diagnóstico, edad, valores de laboratorio de la presentación inicial y la escala de actividad de vasculitis de Birmingham confirmados con biopsia renal (ANCA positivo) identificados en un hospital de especialidades de Ecuador.

Palabras clave: vasculitis, ANCA, anciano, granulomatosis Wegener, enfermedad glomerular, lesión renal.

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Abstract

Currently we live a process of transition in the population pyramid. Therefore, kidney diseases become more frequent in the elderly. Thus, primary vasculitis in older adults are rare diseases, caused by the inflammation of blood vessels and poorly diagnosed. The literature is limited to few clinical cases. Cytoplasmic anti-neutrophil antibody (ANCA) positive vasculitis (AAV) is more common in people older than 50 years. The detection of ANCA gives a very poor prognosis in this population. Also, high suspicion and timely diagnosis is important, as this can provide optimal treatment and reduce the complications of the disease and those associated with immunosuppression. Older adults with AAV have greater renal involvement, hypertension, dyslipidemia and increased mortality. Therefore, two clinical cases from the date of diagnosis, age, laboratory values of the initial presentation and the scale of activity of Birmingham vasculitis confirmed with renal biopsy (ANCA positive) identified in a specialty hospital of Ecuador are detailed below.

Key words: ANCA, vasculitis, elderly, microscopic polyangiitis, glomerular disease, renal impairment.

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Introduction

Vasculitis are divided into two categories: primary vasculitis syndrome and secondary vasculitis syndrome[1]. Primary vasculitis in older adults are caused by inflammation of the blood vessels and are rare diseases[2]. According to the 2012 Chapel Hill Consensus Conference, vasculitis are classified according to the size of the affected vessel and to the ANCA positivity (AAV) [3]. Thus, cytoplasmic anti-neutrophil antibody (ANCA) positive vasculitis is more frequent in people older than 50 years, average age: 62.9 years, annual incidence: 18.1/million,[4] and increases with age.[1] It is characterized by inflammation of the blood vessels[5]. It is defined as a necrotizing vasculitis with few or no immune deposits, predominantly affecting small blood vessels (capillaries, venules and arterioles)[3].

The detection of ANCA is of worse prognosis in this age population[6]. Older adults with AAV have greater renal involvement, hypertension, dyslipidemia and increased mortality[7]. Its clinical consequences are nonspecific, but fever of unknown origin, unintentional weight loss, weakness and polyarthralgia stand out.[1] The literature is limited to few clinical cases. Two cases of young-old patients with a diagnosis of positive ANCA vasculitis confirmed by renal biopsy are described below,

Presentation of case 1

A 67-year-old male patient, born in Catacocha (Loja Ecuador), resident in Quito, (Ecuador), marital status: married, occupation: First Sergeant in the Army in passive service, right-handed, Catholic, blood group: O +, without known allergies. Patient with a history of 4 years of hypothyroidism in treatment with levothyroxine, hyperuricemia in treatment with allopurinol, lung cancer CS III (pt4nxmx) with segmentectomy of the right middle lobe; anatomic pathology report: compatible with adenocarcinoma, moderately differentiated, infiltrative ulcerated positive for malignancy, with two reviews of slides outside of the institution, that indicate infectious process compatible with bronchitis, in 2014.

The patient presented a clinical picture of hyporexia of 8 days of evolution, with no apparent cause, so he went to the first level of care where they found him a rise in blood pressure of 180/95 mm Hg, and administered him captopril 25 mg. without achieving adequate control, they referred him to the specialty hospital, where after assessment, they requested him paraclinical tests finding elevation of nitrogen compounds.

He entered with the following vital signs: blood pressure 168/87 mmHg, heart rate 71 beats per minute, respiratory rate 18 breaths per minute, temperature 36.2°C; conscious patient, oriented in time, space and person, afebrile. On physical examination only a mild edema in lower limbs stood out.

Evolution: the patient was admitted on March 26, 2018 with the following values: hemoglobin 8.5 grs; hematocrit 26.4; platelets 203,000; urea 277; creatinine 19.7; uric acid 12.2; potassium 7.1; sodium 131; chloride 104; cholesterol 142; triglycerides 105; LDL 80; GOT 8; GPT 13; LDH 193; proteins 5.5; albumin 2.4, iron 41; Calcium: 8.4; P: 3.9; sodium: 130; potassium 7.6; TSH: 9.37; FT4 1.25; MU: pH 6; proteins 1+; leucocytes negative; blood 3+; bacteria +; urine sodium 34; creatinine in urine: 133; urine osmolality 600; FeNa: 3.6 %, arterial blood gases: pH 7; 39 HCO3 14.8; pCO2: 24.6; pO2 68.7; lactate 0.7.

It is an acute kidney injury on a chronic kidney disease (Figure 1), because when the renal function history of previous months was reviewed, urea values of 46 and creatinine of 1.43 were observed in December 2018 and this range was maintained until March 2018, a period in which due to the sudden increase in the values of nitrogen compounds, it was suspected a rapidly progressive glomerulonephritis and immunological markers were requested, finding positive P-ANCA (100 u/ml). The tumor markers requested were negative.

Due to these Results, it was suspected a small-vessel vasculitis and anti-MBG antibodies were requested, a renal biopsy was carried out with the following microscopic report: renal parenchyma

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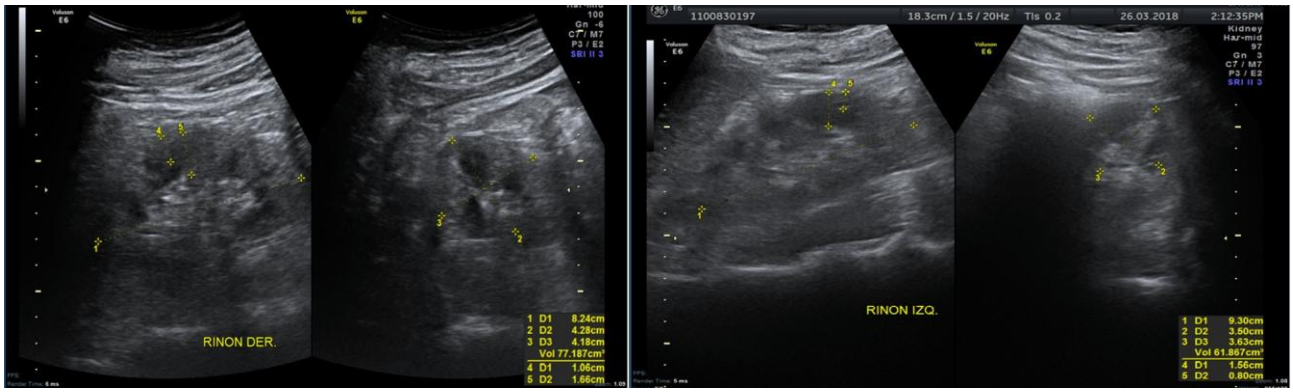

Figure 1. Renal ultrasound: kidneys of usual shape, size and situation, increased echogenicity; no focal lesions are seen, altered cortico-medullary relationship. Pyelocalycial dilation is not observed. The right kidney measures 8.2 x 4.2 x 4.1 cm, parenchyma 16 mm, cortex 10 mm. The left kidney measures 9.3 x 3.6 x 3.5 cm, parenchyma 15 mm, cortex 8 mm. Bladder in scarce repletion, endoluminal lesions are not seen. Scarce liquid in Morison, approximate volume of 5 cc.

showing cortex and medulla, with 4 glomeruli per section in total, extracapillary proliferative lesions: three of active type and one fibrocellular. Disperse interstitium, inflammatory leukocyte infiltration and some neutrophils. Blood residues and occasional calcium oxalate crystals can be observed in the lumen of some tubules. In addition, there is cytoplasmic vacuolization of the tubular lining epithelium, and reparative changes. Interstitial fibrosis associated with tubular atrophy in 10%. The interstitial arteriolar vessels have mild thickening of the wall due to fibrosis. Immunofluorescence: negative IgG, IgA, IgM, C3 and fibrinogen. Diagnosis of active proliferative extracapillary glomerulonephritis of pauci-immune type, suggestive of small-vessel vasculitis with renal involvement.

Treatment includes corticotherapy (pulse of methylprednisolone 500 mg IV for 3 days and then 1 mg per kg of body weight orally) and renal replacement therapy by hemodialysis. The Results of anti-basement membrane tests were negative, and due to high suspicion of small vessel vasculitis, 7 plasmapheresis sessions were added.

The patient was discharged in better conditions due to the stabilization of the clinical picture and hemodialysis treatment and de-escalated doses of prednisone are continued until now without pulmonary complications.

Presentation of case 2

A 67-year-old male patient, mestizo, born in Guayaquil, resident in Quito-Ecuador, passive military, Jehova's witness. Without personal pathological antecedents, no more relevant data. The patient was admitted to the specialty Hospital due to a clinical picture of constitutional syndrome, characterized by nocturnal diaphoresis, fever, cough without expectoration, rhinorrhea and involuntary weight loss (8 kilograms) of 3 months. On physical examination, the patient entered in fair general conditions, with the following vital signs: blood pressure 130/80 mmHg, heart rate of 80 beats per minute, 20 breaths per minute, temperature 38.5°C, oxygen saturation 90 % at ambient air. No positive findings on general inspection.

At the beginning, the service of Internal Medicine requested paraclinical tests, which showed leukocytosis and neutrophilia, creatinine 2.03; urea 65. CT scan of paranasal sinuses without occupation. Bronchoscopy with epithelial atypia. Negative induced sputum culture. Renal ultrasound with preserved morphology, echogenicity and size of both kidneys, bilateral renal lithiasis with left predominance, left renal cyst and extrarenal pyelectasis. Chest CT scan, right axillary lymph node, thickening of the ileocecal wall of 12 mm. Echocardiogram: grade II hypertensive heart disease, normal left ventricular (LV) systolic

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function and preserved in right ventricle at rest. Moderate diastolic dysfunction, with increased LV pressures at rest. Carpentier type 1 moderate mitral regurgitation. Normal pulmonary pressures No vegetations in heart valves.

Taking into account that the patient had no pathological antecedents and due to the elevation of nitrogen compounds, it was requested an assessment by the nephrology service. Previous studies were gathered, in 2017 positive P-ANCA. Because of the epidemiology, the clinical picture and the non-identified cause of acute kidney injury, it was requested a immunological profile that reported negative ANA, ANTI DNA SS and DS, anti B2 GP1, ANCA-C, cardiolipin, ANTI SMITH, antilupus and anti-basement membrane antibodies, and positive P-ANCA. Negative viral studies. Renal biopsy (Figure 2). Therefore, treatment was started with pulses of methylprednisolone and alkylating agents

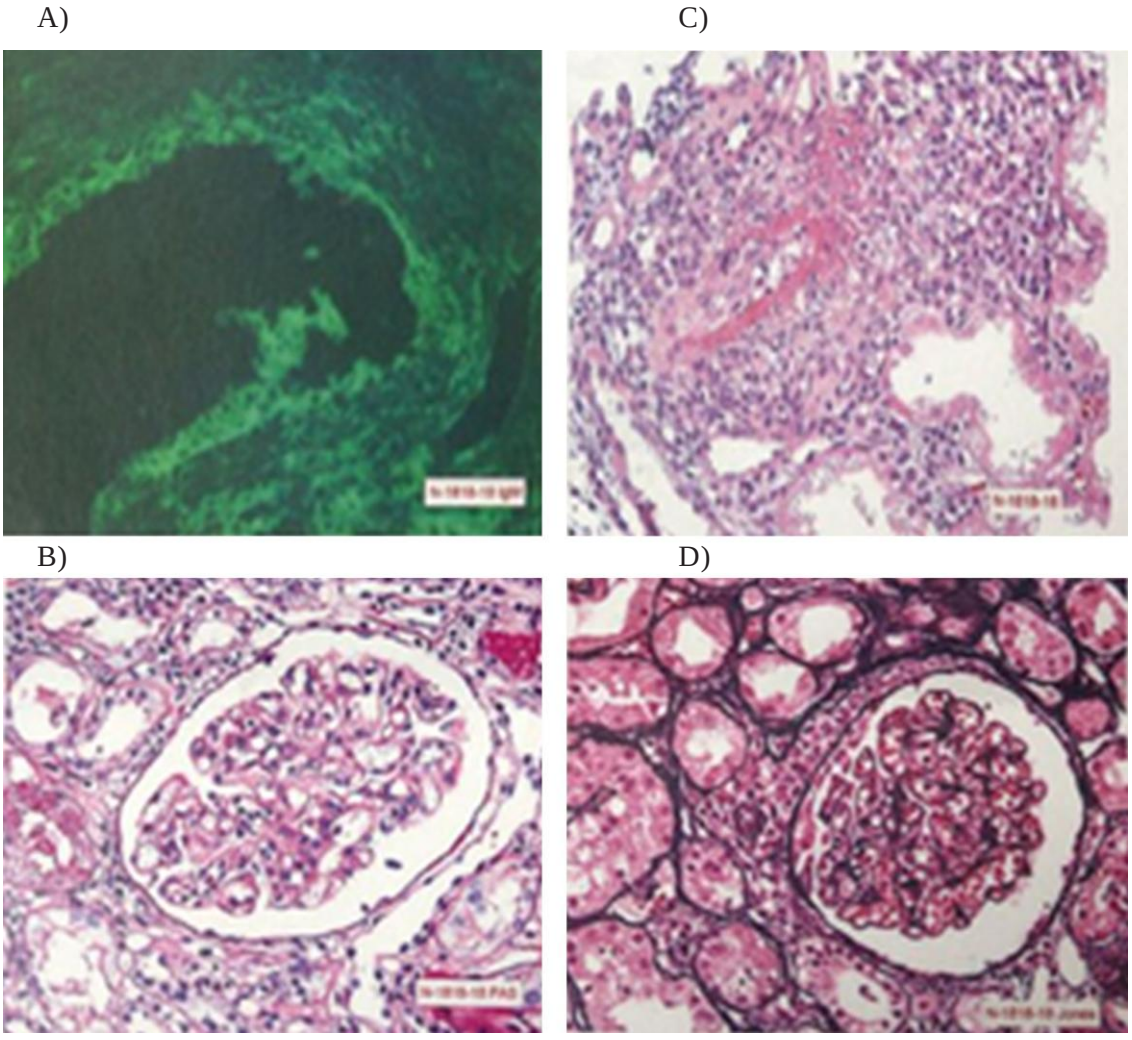

Figure 2. A) Direct immunofluorescence: Negative IgG, C3 and C1q. IgA scarce granular deposits in the wall of some vessels. IgM fine granular deposits in the wall of some vessels. Fibrinogen: interstitial fibrillar deposits. B) Glomerulus hematoxylin-eosin staining. C) In the interstitium there are medium and large caliber vessels with hyalinization, fragmentation of the elastic layer and infiltration of polymorphonuclear cells and lymphocytes that sometimes advances to the tubules and the peripheral interstitial tissue. D) Mesangial hypercellularity. Conclusion: hystopathological image compatible with systemic vasculitis.

(cyclophosphamide). Despite the therapy established, the patient presented hemoptysis and tachypnea, and for this reason was admitted to an intensive care unit with a Birmingham vasculitis activity score (BVAS) (ICU) of 19 points (high risk of mortality) and a radiograph suggestive of pulmonary congestion (Figure 3).

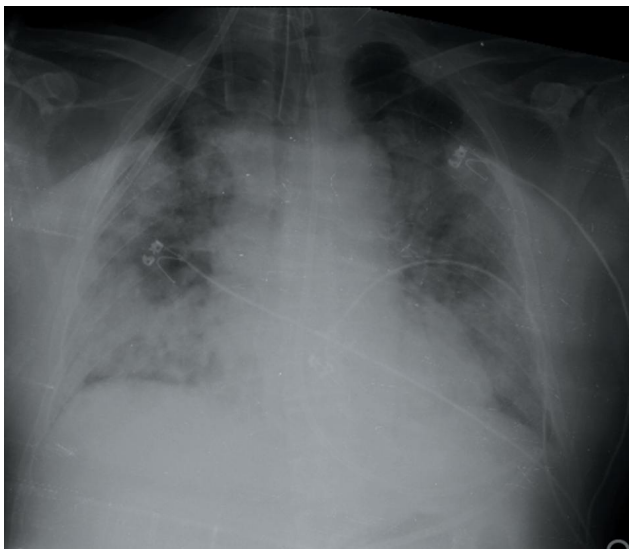

Figure 3. Chest X-ray 20/5/2018.

In the intensive care unit he was prescribed 7 sessions of plasmapheresis, together with peritoneal dialysis. However, the patient continued with a complication of infectious type, and therefore, the prescription of rituximab was not possible. The patient presented multi-organ failure due to sepsis and died.

Discussion

With the change in the population pyramid, increased aging and life expectancy, vasculitis are more frequent in the elderly with a poor prognosis in the short term, since approximately 50% of mortality occurs in the first month after diagnosis. Of the other 50%, only 5% recover renal function. Most patients need renal replacement therapy. Therefore, high suspicion and timely diagnosis are important, to be able to provide optimal treatment and reduce the complications of the disease and those associated with immunosuppression. In this type of patients, it should be carry out a complete evaluation for the identification of hidden infections (bacterial, viral, fungal and parasitic), prophylaxis for opportunistic infections and finally, the prescription of optimal doses of individualized immunosuppression.

Conflict of interest and Funding

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Ethical responsibilities

Protection of people and animals

The authors declare that no experiments were performed on human beings or animals for this research.

Data confidentiality

The authors declare that they have followed the protocols of their workplace on the publication of patient data.

Right to privacy and informed consent

The authors declare that patient data do not appear in this article

Contribution of the authors

In the elaboration of this article all the authors have contributed in the process of selection, writing and edition thereof.

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