Crisis renal esclerodérmica: una causa rara de hipertensión potencialmente mortal

Scleroderma renal crisis: a rare life-threatening cause of hypertension

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ABSTRACT

Scleroderma renal crisis is a rare and severe complication of systemic sclerosis. There are many risk predictors for the development of this complication. Diagnostic criteria are non-consensual, including arterial hypertension and oliguric acute kidney injury (AKI). Angiotensin-converting enzyme inhibitors are an effective treatment, with huge impact in prognosis.

We describe a case of a scleroderma renal crisis in a 78-years-old woman who was diagnosed with systemic sclerosis and treated with prednisone at high dosage, who presented with new onset congestive heart failure, arterial hypertension and oliguric AKI. The diagnosis of scleroderma renal crisis was performed, and angiotensin-converting enzyme inhibitor was initiated with blood pressure control and slightly improved renal function.

The prognosis of scleroderma renal crisis remains poor with high 5-year mortality rate. Medical awareness on tensional values and well-known risk factors could have a huge impact in diagnosis and prognosis and even on prevention of this complication.

Keywords: Hypertension; Acute Kidney Injury; Scleroderma renal crisis.

Palabras clave: Hipertensión; Insuficiencia Renal Aguda; Crisis Renal Esclerodérmica.

BACKGROUND

Scleroderma renal crisis (SRC) is a rare life-threatening complication of systemic sclerosis (SS), characterized by accelerated arterial hypertension of new on set and rapidly progressive oliguric acute kidney injury (AKI). Although incidence has fallen, SRC remains a relevant cause of mortality and its prompt diagnosis and treatment has a significant impact in patient outcome[1-3].

CASE REPORT

We present the case of a 78-years-old women with history of dyslipidaemia, ischemic leukoencephalopathy, colonic diverticulosis and diffuse SS diagnosed 2 years before, presented with skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints; digital tip ulcers; telangiectasias; raynaud's phenomenon and anticentromere antibodies (total 19 points of *ACR/EULAR* criteria). She was chronically medicated with long-term low daily dosage of glucocorticoids that was increased, 2 weeks before hospital admission, to 20 mg/day due to an assumed flare of SS.

She referred dyspnea and lower limb oedema during precious days. On hospital admission, neurologic examination was similar to usual status; corporal temperature was 36.4ºC; pulse was 88 beats per minute; and blood pressure was high (170 mmHg systolic and 90mmHg diastolic maximum pressures). She had inspiratory crackles in lower half of both lungs at pulmonary auscultation, symmetric lower limb oedema and oliguria. Laboratory analyses showed AKI (basal serum creatinine 0.8mg/dL increased to 3,4mg/dL on admission) without anaemia or thrombocytopenia, with normal values of bilirubinaemia and haptoglobin. Blood gas analyses showed no metabolic acidaemia or hyperkalaemia. Anti-nuclear antibodies were positive (1/1280 – homogeneous pattern) with no complement consumption and anti-RNA polymerase III absent. Urinalysis revealed haematuria (200 mg/dL) and non-nephrotic proteinuria (0,64 g/daily) without leukocyturia or dysmorphic erythrocytes. Renal and bladder ultrasound showed normal kidneys and excluded hydronephrosis or other complications. Transthoracic echocardiography documented new moderate to severe left ventricular systolic dysfunction and mild pulmonary hypertension (pulmonary arterial pressure +/- 40mmHg). Coronariography ruled out coronary disease. Diuretic treatment was initiated on the admission day, with significant improvement of hypervolemic symptoms but, however, maintaining severe arterial hypertension along with progressive worsening of AKI.

Due to known history of systemic sclerosis and presence of multiple risk factors (Table 1), SRC was diagnosed, and angiotensin-converting enzyme inhibitor was initiated. Captopril dosage was progressively adjusted to maximum daily dosage tolerable, and blood pressure control was achieved with a slightly improve in renal function and diuresis. Renal replacement therapy was not required, and renal biopsy was not performed. Although clinical improvement of SRC, latter on hospitalization, the patient died due to a nosocomial sepsis.

DISCUSSION AND CONCLUSIONS

SS is a rare systemic autoimmune disease with fibrosis in skin and internal organs, including the kidneys, heart, lungs and gastrointestinal tract[1,4]. SRC is the most important re-

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Table 1. Risk predictors of Scleroderma Renal Crisis

Predictors of Scleroderma Renal Crisis [1,2,4,7,8]
- Diffuse skin involvement with rapid progression,
- rapid skin thickening,
- systemic sclerosis duration of less than 4 years,
- new onset anaemia,
- cardiac events (including pericarditis and congestive heart failure),
- arthralgias, synovitis, tendon friction rubs, large join contractures,
- positive test for anti-RNA polymerase III antibodies,
- corticosteroid of an equivalent dosage of prednisone higher than 15-20mg/day,
- female sex,
- African Americans,
- exposure to cold climate

Table 2. Diagnostic Criteria os Scleroderma Renal Crisis

Hypertension Criteria
>140/90 mmHg in previous normotensive patients
or
>30mmHg rise in baseline systolic blood pressure
and
>20mmHg rise in diastolic blood pressure)[3]
Renal Failure Criteria
≥ 50% serum creatinine increase over patient's baseline
or
≥ 120% serum creatinine over upper limit of normal[1]

nal complication of SS[5]. Although its pathogenic mechanisms are not completely elucidated, it has been affiliated to a decreased renal cortical blood flow caused by intimal thickening of renal interlobular and arcuate arteries due to several potential hypothesis: endothelial cell injury, platelet factors release with increased vascular permeability, fibrin deposition and collagen formation and episodic renal vasospasm such "renal raynaud". Renal hypoperfusion induces increased production and release of renin causing malignant hypertension[1,5,6].

As a rare disease, it stands for only 5% of patients with diffuse cutaneous form of SS with current fallen incidence, due probably to widespread use of vasodilators and more careful use of corticosteroids[3].

Although absence of consensual diagnosis criteria of SRC, it is characterised by rapid and sudden increase in arterial pressure and rapidly progressive oliguric AKI (Table 2). Other manifestations such malaise, fatigue, headache, seizures, fever, encephalopathy, blurred vision, dyspnoea, pulmonary edema, arrhythmias, myocarditis and pericarditis can be present. Laboratory common findings that could prove the diagnosis are the increase in serum creatinine level with mild proteinuria and microscopic haematuria (hemoglobinuria in most cases) and thrombotic microangiopathy with haemolytic anaemia and thrombocytopenia, present in half of the cases[1,2,3,6,7].

Kidney biopsy could support but is not required to make SRC diagnosis. Typical histologic features in renal biopsy are the predominance of small vessel involvement with thrombi, mucoid changes and concentric intimal fibroplasia with "onion-skin" in interlobular artery, perivascular fibrosis and ischemic glomerulus[1,6].

There are well-studied clinical predictable factors linked to a higher risk of developing SRC (Table 1). In our clinical case the female gender, short disease duration, use of high dosage of prednisone and the acute cardiac event such as congestive heart failure were the risk predictors identified[1,2,4,6,8].

SRC treatment remains adequate with the use of angiotensin-converting enzyme inhibitors to control blood pressure and optimize renal perfusion. Captopril is the preferred one, due to its rapid onset of action and short half-life. If blood pressure remains not controlled with maximum dosages of captopril, calcium-channel blockers and diuretic or al-

pha-blockers should be added, by this order, until optimal results. Dialysis is committed to acute setting scleroderma renal crisis – associated uremia despite appropriate medical therapy and in end-stage kidney disease. Despite being associated to a decreased survival, renal function recovery happens in approximately 40-50% of the patients submitted to dialysis therapy. Due to this findings, renal transplantation should be delayed at least 12-18 months after dialysis initiation[1,3,7,8,9]. Despite the dramatically reduced mortality since ACE inhibitors use, SCR prognosis is still poor with a 5-year mortality rate about 50-70%[3]

Prevention with monitoring blood pressure in those patients at high risk of developing SRC plays a key role in early diagnosis. Although this, there is not a prophylactic therapy regarding to this population[1].

The main purpose of this clinical case is to improve medical awareness of SRC as a rare cause of hypertension in patients with known SS. Despite there are no preventive measures available, consciousness of this disease could generate more surveillance in patients that are in risk of SRC.

CONFLICTO DE INTERESES Y FUENTES DE FINANCIACIÓN

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