Kidney biopsy is mandatory in cases of silent arterial hypertension in scleroderma renal crisis: a case report

Sir,

Scleroderma renal crisis is classically characterised by a rapid and progressive renal failure associated with malignant hypertension, and occurs in 5–10% of the patients with diffuse systemic sclerosis (SSc) (1-3). Therapy with angiotensin-converting enzyme (ACE) inhibitors is mandatory, leading to stabilisation of the renal failure, decreasing mortality significantly, from 76% to 15% in the first year (4-6).

Nevertheless, it has been described some “silent” cases, without the characteristic malignant hypertension. This atypical clinical presentation can delay the diagnosis, and consequently the treatment with ACE-inhibitors, leading to an increased mortality (3, 7).

In December 2008, a 45-year-old woman female with diffuse SSC was attended in our institution. She was previously treated with prednisone 40 mg/day, without improvement. The BP was 130 x 90 mmHg and laboratory tests were normal, with a positive antinuclear antibody (ANA) (1/160, speckled pattern). She was prescribed cyclophosphamide and prednisone was tapered. After 17 months infusions, she presented hemorrhagic cystitis, and cyclophosphamide had to be substituted by azathioprine 150 mg/day. Her BP stayed at a normal range during the entire treatment.

In May 2011, the patient presented a rapid decrease in renal function (creatinine ≥ 4.66 mg/dL), associated with BP of 150 x 90 mmHg, and was hospitalised. Lab tests revealed no haemolytic anaemia, plaque-topenia, proteinuria or haematuria, with negative anti-dsDNA and ANCA, and positive anti-RNA polymerase III. Renal Doppler ultrasonography showed no stenosis of renal arteries. Although the patient normalised BP with captopril 100 mg/day, the progressive renal failure continued, and haemodialysis was initiated. A kidney biopsy was performed, confirming SRC (Fig. 1).

The presence of obliterating endarteritis with onionskin appearance, narrowing of arterioles and glomerular ischaemia, without inflammatory changes or immune deposits, are characteristics of SRC (5). For diagnostic purpose in this patient presented here, kidney biopsy was fundamental to establish the diagnosis of SRC. It also helped to distinguish SRC from ANCA-associated glomerulonephritis, which has been reported in SSC, and should be treated with high doses of steroids and immunosuppression (6). The early diagnosis and the subsequent start of the therapy with ACE-inhibitors are key for the success of the treatment of SRC.

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