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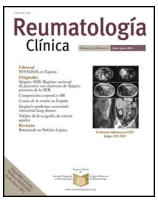

Letter to the Editor

Scleroderma renal crisis: The experience of a third-level hospital☆

Crisis renal esclerodérmica: la experiencia de un hospital de tercer nivel

Dear Editor,

Systemic sclerosis (SS) scleroderma is a systemic autoimmune disease with excessive deposits of collagen, vascular lesions, inflammation, fibrosis of the skin and of different internal organs.[1–3]

The severest renal involvement is the scleroderma renal crisis (SRC), a rare complication with a prevalence of 4%–6%: 7%–9% diffuse SS and .5%–.6% limited form SS.[4]

In SRC new onset accelerated high blood pressure occurs, and rapid progression oliguric renal failure in the context of SS. Between 11% and 14%, of patients have normal blood pressure levels.[2,3,5,6]

We present a series of 5 cases of SRC who were cared for in our hospital and describe the main clinical characteristics, immunological profile, and disease evolution.

Five cases were identified out of a total of 150 patients diagnosed with SS in the University Hospital Central de Asturias from 2000 until 2017. Their characteristics are contained in Table 1. Diagnosis of SRC was made in according with the criteria proposed by Penn et al. in 2007.

Determination of antinuclear antibodies (ANA), anti-topoisomerase (Scl-70) and anticentromere antibodies were made using immunofluorescence.

Of the 5 cases there were 4 diffuse SS and one SS sine scleroderma. Eighty per cent were women with a median age of 59 years. In 3 patients diagnosis of SS was established with the initiation of SRC and in 2 cases this occurred between the first signs of SS and the diagnosis of SRC was at 3 and 48 months, respectively. In one case BP values were normal, the others presented with accelerated high blood pressure. One hundred per cent had renal failure. In the immunological study the ANA were positive in 5 patients. The Scl-70 were positive in 3 patients and in 2 the anticentromere was positive.

One case required diagnostic renal biopsy. Forty per cent required haemodialysis; 2 died. During the SRC they all received treatment with the angiotensin-converting enzyme inhibitors (ACE).

Table 1
Characteristics and evolution of the 5 patients with Scleroderma Renal Crisis.

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Age	59	47	53	60	78
Sex	Female	Female	Male	Female	Female
Time since diagnosis (months)	48	0	4	0	0
Type of scleroderma	Diffuse	Diffuse	Diffuse	Sine scleroderma	Diffuse
Clinical symptoms					
DILD	Yes	Yes	Yes	No	Yes
HBP previously	Yes	No	No	No	No
Previous anti-hypertensive treatment	ACE	–	–	–	–
HBP initially	Yes	Yes	No	Yes	Yes
Raynaud	Yes	No	No	Yes	Yes
PHT	Yes	Yes	No	No	Yes
LHF	Yes	No	Yes	Yes	Yes
Immunological profile					
ANA/pattern	1/1280 Homogenous	1/320 Mottled	1/2560 Homogeneous + nucleolar	1/2560 Centrometric	1/1280 Homogeneous
Anti-Scl-70	+	–	+	–	+
Anticentromere	–	+	–	+	–
Evolution					
EGF/ERC	EGF 20	EGF 63	EGF < 15	EGF 17	EGF < 15
Haemodialysis	No	No	Si	No	Si
Prognostic	Died	CRD	Died	CRD	ERC
Possible precipitants	Undetermined	Undetermined	Undetermined	Undetermined	Prednisone: >7.5 mg

ANA: antinuclear antibodies; DILD: diffuse interstitial lung disease; CRD: chronic renal; FGE: estimated glomerular filtration; HBP: high blood pressure; PHT: pulmonary hypertension; ICI: left heart failure; ACE: angiotensin converting enzyme inhibitors; Scl-70: anti-topoisomerase antibodies.

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SRC is a potentially mortal complication with a prevalence of 3.3% in our hospital, more frequently at the start of the diffuse forms, and is able to present on diagnosis, as occurred in 3 patients of our series where the SRC was diagnosed from SS.

Risk factors predicting SRC have been described as: anemia, recent cardiac involvement, anti-RNA polimerase III antibodies and the use of corticoids (>15–20 mg/day).[1,2,5] In the majority of our patients no possible trigger was identified. One of our patients had been in treatment with corticoids at doses above 7.5 mg/day.

There are no data to show that pre-existing factors such as high blood pressure, proteinuria, raised creatinine levels, anti Scl-70, anticentromere antibody, or previous histological renal changes are associated with a higher frequency in the development of the SRC.[2]

Morbimortality by SRC is high and difficult to manage. It is a medical emergency, initially focused on BP control. ACEi are the drugs of choice, even in patients with renal failure. There is controversy regarding its prophylactic use as previous treatment with ACEi prior to the diagnosis of SRC may obscure its presentation.[7]

Dual therapy: ACEi and the endothelin inhibitor (bosentan) have been used in a series of 6 patients, with a follow-up of 5 months with improvement of glomerular filtration, with no changes in mortality and without studies to endorse its safety.[7] Sixty per cent of patients will require renal replacement therapy.[2,6]

When there are signs of accelerated blood pressure with oliguric renal failure sometimes accompanied by thrombotic microangiopathy of unexplained causes and cardiological involvement (pericarditis, cardiac blockage) we must direct out investigations to the diagnosis of an SS, usually diffuse, since SRC is a treatable complication if there is early identification.

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Rheumatoid arthritis patient preferences for the treatment administration route☆

Preferencias en la vía de administración del tratamiento de pacientes con artritis reumatoide

Dear Editor,

Treatment for rheumatoid arthritis (RA) has advanced greatly over the last 20 years with the incorporation of biologic therapies to the therapeutic arsenal the rheumatologist has at his or her disposal. Initial biologic intravenous (IV) treatments led to a large number of subcutaneous (SC) drugs, and in recent years new non biologic oral drugs have come onto the market. Up until now the patient had little say regarding the ideal administration route, but this has recently changed.[1–3] Patient opinion is increasingly more important in choosing the mechanism of action of a biologic, and also its form of administration.[4]

In order to determine the current opinion of a sample of patients with rheumatoid arthritis (RA) attended in our centre all patients with RA who used the rheumatology service were successfully selected for 2 weeks. Each patient was asked 3 questions by the nursing staff (day hospital and biologic therapy unit) or by their regular rheumatologist (outpatient consultations). The administration route of the current treatment of each patient was also recorded, forming patient groups with oral administration (only oral treatment), subcutaneous administration (subcutaneous treatment with or without oral treatment) and intravenous treatment (with or without oral treatment). The first question was: what is the ideal administration route for you in a RA treatment? The second was: Why did you choose this administration route? And the last was: did you at any time talk about the administration route with the rheumatologist in charge of your treatment? Response options patients could give for the first questions were 3, oral, subcutaneous or intravenous administration route and for the third question were 2, yes or no. The second question allowed for more of an open response which was summarised in the options contained in Table 1. Those patients who had responded to the questions in the day hospital or the functional unit of biologics were excluded when they were attended by outpatient departments.

Overall patient responses are contained in Table 1. The patients with oral treatment received methotrexate (7 patients) leflunomide (4 patients) and azathioprine (one patient); the patients with subcutaneous treatment received etanercept (6 patients), adalimumab (4 patients), golimumab (4 patients), certolizumab (2 patients), abatacept (one patient), tocilizumab (3 patients) and methotrexate (4 patients); and the patients with intravenous treatment received infliximab (23 patients), tocilizumab (18 patients), rituximab (11 patients) and abatacept (5 patients). Those patients with oral treatments were content with the oral administration route (10/12), whilst the majority of patients in SC treatment would have preferred an oral drug (16/24 patients) due to its convenience (69%). With regard to the patients treated with intravenous therapies, surprisingly, the majority (44/57) preferred the IV route, in contrast to that expressed in several published studies.[2] The good

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