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Case Report Open Access

Scleroderma Renal Crisis without Cutaneous Involvement: One Report Case in a Black African

Eliane Mikkelsen Ranivoharisoa[1*], Benja Ramilitiana[1], Olivah Henintsoa Rakotonirainy[2], Harinjara Razanakoto[2], Fahafahantsoa Rapelanoro Rabenja[2] and Willy Franck Harilalaina Randriamarotia[1]

[1]Department of Nephrology and Hemodialysis, Befelatanana University School of Medecine, Antananarivo, Madagascar

[2]Department of Infectious Disease, Befelatanana University School of Medecine, Antananarivo, Madagascar

[*]**Corresponding author:** Ranivoharisoa EM, Department of Nephrology and Hemodialysis, Befelatanana University School of Medecine, Antananarivo, Madagascar, Tel: 261341420161; E-mail: remikmed@hotmail.fr

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Abstract

Objective: To report an atypical case of renal scleroderma crisis without cutaneous involvement and no serological manifestation.

Observation: A black African, 33 years old was hospitalized for a malignant hypertension. He reached rapidly the end-stage of chronic kidney disease due to a scleroderma renal crisis. Presumption diagnostic was established by clinical signs, biological and imagery investigations.

Results: The patient didn't have personal and familial medical backgrounds a part losing 20 kilos two months before hospitalisation. In admission, he has malignant hypertension 220/140 mmHg and there was no cutaneous involvement of scleroderma. Blood tests concluded a chronic kidney disease with an estimated Glomerular Filtration Rate less than 2 ml/mn/1.73 m2. The C- reactive protein was negative and the sedimentation rate in first hour was accelerated to 119 mm. The proteinuria was 0.8 g/24 hours. The immune balance of the ribonucleic acid polymerase, the anticytoplasmic antibodies, the antinuclear antibodies were repeated more times but were negative. Imagery investigations found systemic manifestation such as lung fibrotic with atelectasia. Heart doppler found hypertension in pulmonary artery and minim pericarditis. Renal Doppler confirmed hypotrophic sizes of both kidneys with high index resistances in small vessels. Renal biopsy was not done. Six months later appeared limit digitals ulceration on both fingers. The patient was treated by angiotensin-converting-enzyme inhibitor, calcic inhibitor and underwent to a chronic hemodialysis with significant improvement mainly about the blood pressure (120/70 mmHg).

Conclusion: Any malignant hypertension in scleroderma in the absence of other causes with a rapid deterioration of renal function should think first of all a scleroderma renal crisis, cutaneous involvement and serological manifestation are not always obligatory.

Keywords: Chronic kidney disease; Scleroderma renal crisis; Systemic scleroderma; Malignant hypertension; Angiotensin-converting-enzyme inhibitor; Dialysis

Introduction

Scleroderma or systemic sclerosis is defined as a connectivity tissue disorder which involves skin, gastrointestinal tract, kidneys and lungs. Scleroderma renal crisis (SRC) is characterized by the presence of high blood pressure (HBP) associated with oligoanuria, and renal dysfunction. It is the most severe and sometimes fatal complication of scleroderma. In nephrological field, it is always considered as an emergency therapeutic [1]. In the absence of an early treatment, patient can lose definitely his renal function.

According to our acknowledges, SRC without cutaneous manifestation is infrequent and few observations have been reported worldwide. U Zwetter in his observation has reported one atypical case of SRC. In the admission at hospital, there was unremarkable skin lesion but followed lately by the development of several skin manifestation such as telangiectasia, sclerodactyly, and finished by generalized cutaneous sclerosis [2]. In Gonzalez's report case, he described one atypical SRC with cutaneous involvement limited on fingers and feet but no serological manifestation repeated more times in an old man [3]. Our objective is to report another case of SRC in a black African (Malagasy) associating the absence of skin lesion in admission without a serological manifestation. Six months later appeared limited digital ulceration on both fingers.

Case Presentation

A 33-year-old man, Malagasy was hospitalized in rheumatology department for a management of a malignant hypertension.

His medical history begun two months before his admission by physical asthenia complicated by a weight losing of 20 kg. He didn't have transit disorder or dysphagia or fever. He had no specific personnel and familial backgrounds a part smoking cigarettes with 5 packages per year.

In admission, patient has a malignant hypertension (220/140 mmHg). He complained about headache scored 4/10 according to digital scale, vomits and epigastric pain.

He had neither seizures nor neurological deficits. Physical examination concluded signs of retinopathy hypertensive staged III, regular sinusal tachycardia, double sound B2 on hearth auscultation, bilateral pleuretic syndrome and oedema on lower limbs. The abdomen examination was normal with any palpable mass. There were no identifiable cutaneous manifestations of scleroderma like sclerodactyly or telangiectasia.

In paraclinical signs, the blood count showed normochrom normocytic anemia with 10 g/dl of hemoglobin. Lactate dehydrogenases and haptoglobinemia were in theirs normal ranges. The sedimentation rate was accelerated in 119 mm at the first hour; C-Reactive Proteine was flat. The ionogram showed chronic hypokalaemia. The serum creatinine was 831µmol/l equivalent of an estimated glomerular filtration rate 2 ml/min/1.73 m^2 according to chronic kidney disease epidemiology; the urea was 38 mg/dl. Proteinuria was 0.8 g/24 hours. Hepatitis B and C, and HIV serology were negative. Renal ultrasound coupled with Doppler excluded the presence of renal artery stenosis and showed small sizes of kidneys. In intra parenchymatous, there were high index of resistance up 0.83 in the left kidney and up to 0.86 in right kidney, observed mainly in interlobar and segmental arteries. The arched arterieswas not visualised. Pulmonary tomodensitometry revealed an interstitial syndrome, pulmonary fibrosis with an atelectasis and minimal pleurisy. Pleurisy was transudative and no malignant cells were found.

Heart doppler found minimal non-compressive posterior pericarditis and high systolic pulmonary arterial hypertension (51 mmHg). Six months later appeared a limited digital ulceration on both fingers. From the therapeutic point of view, the patient underwent on a periodic dialysis with a medical treatment associated Angiotensin-Converting-Enzyme (ACE) inhibitor, calcic channel blocker. There was an improvement with the achievement of the blood pressure target.

Discussion

Scleroderma is a connectivity disease characterized by inflammation and fibrosis involvement in the skin and other organs including the heart, lungs, digestive tract and kidneys with vascular abnormalities [4]. The SRC is a serious complication, reported firstly in 1863 [5]. It is a vasculitis of small vessels resulting the intimal proliferation of vascular lumen and inducing a decrease renal blood flow [6].

With the development of ACE inhibitors, survival rate has improved dramatically but death rate still remains unacceptably high. It occurs mainly in the early years of the disease in patients with systemic scleroderma and occurs in 2-10% of scleroderma [7]. In some cases, SRC was the initial presentation of scleroderma; there were no skin sclerosis during the time that SRC develops [8]. In this observation, we reported one case of a young Malagasy who reached rapidly the end stage of CKD secondary to a SRC. According to the literature, SRC occurs mainly in men [9]. But other observations also reported its occurrence on women [10]. The average age is variable, ranging from 27 to 75 years. This is an autoimmune disease which etiology was not yet well demonstrated. However, the reported risk factors are mainly diffuse cutaneous manifestations, the black race, the use of high-dose corticosteroids, cold [11,12]. Apart from the black race, we were not able to identify any other favourable factors. Concerning the symptomatology, HBP and renal dysfunction are usually constants, often associated with oligoanuria and micro-angiopathic haemolytic. Blood Pressure is malignant in 75% of cases [13]. In our observation, the patient presented malignant hypertension and its various complications associated with a rapid deterioration of the renal function. In Molina's observation, the patient also presented the same signs with unexplained weight loss during a SRC [14]. The paraclinical examination in our observation concluded anaemia without signs of microangiopathic haemolytics. According to the literature, the signs of thrombotic microangiopathy are not always obligatory, they can be absent in one third of the cases. Concerning the various immune tests, the RNA polymerase, the anti-topoisomerase antibody is particulary specific for the SRC [15]. Another study conducted by Gonzalez also showed the negativity of these antibodies tests even if the controls were carried out several times [3].

As SRC is caused by the vasculitis of small vessels, the echo-doppler has its important place to help for the diagnosis. It shows an increased resistance index mainly in small vessels including inter-lobular and segmental, arched arteries confirming the sites of vascular attacks. A study conducted by Durant et al. referred to the evaluation by arterial echocardiography in 48 sclerodermic patients and concluded his interests [16]. Renal biopsy is not indicated beyond the normalization of blood pressure. In the observation of Najoua, the biopsy was in favour of the signs of vasculitis in the intraparenchymatous arteries including indirect signs of thrombotic microangiopathy [17,18]. In our case, the patient was seen late with end-stage kidney failure with small bilateral kidneys contraindicating the realization of renal biopsy.

In therapeutic view, using ACE inhibitor is recommended in first intention. The indication of hemodialysis depends on each context. According to published cases, most patients underwent for chronic dialysis because of delayed diagnosis [9,17]. Other patients may be weaned in 50% in 24-48 months according to improvement of renal function [19]. According to Penn et al. transplant survival after transplantation remains lower compared to other non-scleroderma patients [20]. In our case, the patient underwent on medical treatment using ACE Inhibitor, Calcic channel blocker with a periodic chronic haemodialysis. There was a good improvement with the achievement of the therapeutic goals (120/70 mmHg).

Conclusions

We have reported an atypical case of a SRC. A young Malagasy of 33 years presented SRC with any cutaneous involvement at the beginning of scleroderma and any serological manifestation but followed six months later digital ulceration on both fingers. The diagnosis was presompted by clinical, biological, imagery signs and therapeutic evolution. The black race, malignant high blood pressure, renal insufficiency and systemic manifestation allowed us to report the diagnosis of a systemic sclerosis with renal manifestation. The patient was seen lately with an advanced stage of chronic kidney disease. He was undergoing on chronic dialysis and his blood pressure was well controlled with ACE inhibitor and calcic channel blocker.

Unexplained malignant hypertensions on systemic scleroderma with a rapid deterioration of renal function associated or no with an oliguria are emergency in nephrology. Despite the absence of skin involvement, SRC should be searched first of all.

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Study concept, interpretation of data drafting of the manuscript: Eliane Mikkelsen Ranivoharisoa, Benja Ramilitiana, Olivah Henintsoa Rakotonirainy, Harinjara Razanakoto.

Critical revision of the manuscript for important intellectual content: Willy Franck Harilalaina Randriamarotia, Fahafahantsoa Rapelanoro Rabenja.

Supervision: Willy Franck Harilalaina Randriamarotia.

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