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Heart Disease Scleroderma Revelators: About a Clinical Case

Coumba Thiam[1], Boubacar Sonfo[1*], Youssouf Camara[1], Asmaou Keita[2], Mariam Sako[3], Békaye Traoré[4], Djeneba Koné[4], Mamadou Touré[5], Massama Konaté[6], Hamidou Oumar Bâ[5], Ibrahima Sangaré[5], Sanoussi Daffe[2], Daouda Fofana[2], Mariam Cheick Traoré[1], Boureima Dembélé[3], Boubacar Diarra[5], Hamidou Camara[5], Samba Sidibé[3], Souleymane Coulibaly[3], Ichaka Menta[5], Ousmane Faye[4]

[1]Department of Cardiology, Kati University Hospital, Kati, Mali
[2]Department of Cardiology, The Mother-Child University Hospital, Bamako, Mali
[3]Department of Cardiology, CHU Point G, Bamako, Mali
[4]Department of Cardiology, The CHU Gabriel Touré, Bamako, Mali
[5]Dermatology Hospital of Bamako, Bamako, Mali
[6]Department of Internal Medicine, Mali Hospital, Bamako, Mali
Email: *sonfo20032001@yahoo.fr

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Abstract

Scleroderma (or systemic sclerosis) is a disease characterized by abnormalities in the functioning of small blood vessels and the immune system, ultimately leading to inflammation and excessive fibrosis of the skin and various organs, including the heart. Management must be multidisciplinary, to avoid complications that are often serious. We report the case of a 20-year-old patient with no known cardiovascular history who consults for dyspnea, and retrosternal pain associated with a dry cough. On physical examination, she had tachycardia, swelling of the lower limbs, jugular turgidity, and deafening heart sounds. Cardiac Doppler ultrasound shows dilation of the right cavities, paradoxical septum and significant pulmonary arterial hypertension, pericardial effusion of medium abundance. On oral examination, it presents an ulceration of the lips, dermatological examination finds scattered hypo chromic spots in the body, more accentuated in the face. Before the hypo chromic dermatosis, a dermatological consultation was carried out with an autoimmune assessment that came back positive for systemic scleroderma.

Keywords

Heart Failure, Scleroderma, Young Subject

1. Introduction

Scleroderma (or systemic sclerosis) is a disease characterized by abnormalities in

the functioning of small blood vessels and the immune system, ultimately leading to inflammation and excessive fibrosis of the skin and various organs, including the heart. Cardiac involvement occurs in 10% to 30% of scleroderma patients. It occurs in both the limited and diffuse forms of the disease, but is more frequent and severe in the diffuse form with skin involvement that progresses rapidly associated with myositis (inflammation of the muscles). Cardiac damage observed in scleroderma patients can be directly related to abnormalities of small vessels, inflammation and fibrosis, or other heart diseases frequently found in the general population aged 50 and over such as atherosclerotic coronary artery disease, valvular pathologies, high blood pressure and its complications [1]. Systemic scleroderma (ScS) is a rare systemic autoimmune disease whose prevalence varies between 50 and 200 cases per million inhabitants, which represents about 10,000 patients in France [2]. Women are more frequently affected than men (sex ratio 8/10) and the disease usually begins between the ages of 40 and 60 [3]. Cardiac involvement of ScS is one of the main severity factors of this disease [4], with a poor prognosis [5], the focus is currently on detecting lesions at the earliest stages, thanks to non-invasive means increasingly accurate, for example ultrasound, cardiac MRI and autoimmune assessment [6]. We report a case of scleroderma, with cardiac involvement in a young subject.

2. Case Presentation

This is a young 20-year-old patient with no known medical and surgical history, gyneco-obstetrically, she is nulliparous. Born of a pregnancy estimated at term, she is the third child of her siblings.

She consults for stage II dyspnea of NYHA, a retro sternal pain associated with a dry cough. The beginning of the symptomatology would go back to about one year marked by a progressive installation of a dermatosis type of speckled hypochromic macula initially sitting on the phalanges of the two upper limbs extending to the face associated with pain of the phalanges and a notion, of generalized pruritus, at the level of the mouth, it presents an ulceration of the lips a fever not quantified. The current episode would go back to about five months marked by retro sternal pain with a type of gravity of gradual installation calmed, by the position leaning forward (Mahometane), or lying down then dyspnea stage II of NYHA, a notion of unquantified fever, puffiness of the face, edema of the lower limbs and cough. Faced with the worsening of these symptoms, the parents decided to take her to the cardiology consultation for better management.

The physical examination had found a weight = 50 kg, height = 159 cm, BMI = 18.5, the general condition preserved with a performance index of the WHO rated at 2, the normo colored conjunctiva, the heart rate at 103 BPM, the blood pressure in the left arm: 100/80mmHg and right arm: 110/80mmHg, oxygen saturation: 98%, temperature: 36.9˚C. There is jugular turgor, the Harzer sign in xiphoid focus, palpation the abdomen was distended with hepatomegaly at 13

cm painful, smooth surface and foam edge, with a jugular hepato reflux, at the lower limbs, we note sock-shaped edema taking the bucket., on auscultation muffled heart sounds, a burst of B2 at the pulmonary focus, fine crackling gratings at the 2 right pulmonary bases.

At the dermatology examination we note hypochromic speckled macules well limited on the back of the hands with respect to the proximal inter phalangial joints (**Figure 1**) but also on the symmetrical internal malleoli and the face associated with proximal and distal inter phalangial sclerosis (PPI), sclerosis of the facial skin with slight taper of the nose and a discreet limitation of the opening of the mouth. A progressive pulpal ulceration of the left major with some pulpal scars.

The neuromuscular examination found muscle weakness (muscle strength 2 - 3) in the arms with a positive scarf sign. A poly arthralgia with pain excuse fingers, elbows and knees without deformation or swelling.

On the basis of these clinical signs, the following diagnostic hypotheses were evoked: autoimmune pericarditis decompensated in right heart failure, complicated pulmonary embolism in right heart failure and primary pulmonary hypertension complicated in right heart failure. After the dermatological consultation the hypothesis of Sharp syndrome associating scleroderma, dermatomyositis and lupus was evoked.

Additional tests were conducted to confirm or confirm the diagnosis. The results were presented in **Table 1** and **Table 2**.

On the electrocardiogram performed we can observe a regular sinus rhythm at 70 BPM, while on the transthoracic cardiac ultrasound (**Figure 2**) we found: dilation of the right cardiac cavities, a paradoxical septum, an important hypertension, systolic function of the RV little altered, systolic function of the conserved LV, pericardial effusion of great abundance (about 25 mm), ejection fraction of the left ventricle (LVEF): 97%, diastolic VD diameter: 43.6 mm, PAPS: 80.4 mmHg, TAPSE:14.4 mm, VCI: 27 mm. Chest tomodensimetry and angiography performed were normal outside a small focus of interstitial syndrome of the left base. Cardiac MRI is currently unavailable at our level. The biopsy not performed

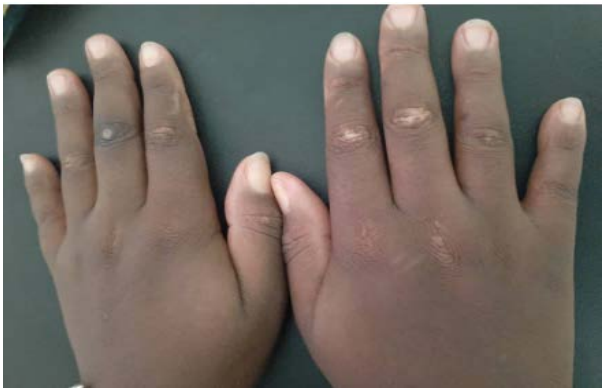

Figure 1. Speckled hypochromic macules well limited localized at the joints proximal interphalagiennes of both hands.

Table 1. Results of paraclinical examinations.

Types of studies	Results
Hemoglobin	11.8 g/dl
Hematocrit	39.7%
Red blood cells	$5.52 \times 10^5/mm^3$
White blood cells	$6.1 \times 10^5/mm^3$
Urea	2.9 mmol/l
Blood creatinine	84 µmol/l
CRP	12 mg/l
Platelets	$346 \times 10^3/mm^3$
Erythrocyte sedimentation rate	1st hour: 3 mm 2nd hour: 9 mm
Transaminases	ASAT: 197 UI/L,
Transaminases	ALT: 187 UI/L
INR	1.54
Proteinuria	0.56 g/24h
Hepatitis B serology test	Negative
Hepatitis C serology test	Negative
Muscle enzyme CPK	228 UI/L

Table 2. The autoimmune balance.

Types of autoimmune tests	Results
Soluble nuclear antibody	76.40 UA/ml (positive)
Anti-SS-A/RO	68 UA/ml (positive)
Anti-SS-B/La	2.90 UA/ml (négative)
Anti-centromer anti-body B	4.30 UA/ml (négative)
AC native anti-DNA	<10.0 UI/ml (négative)
ANTI-MS antibodies	61.6 AU/ml (positive)
Anti-nuclear AC (ANA screen)	2.9 E/S (positive)

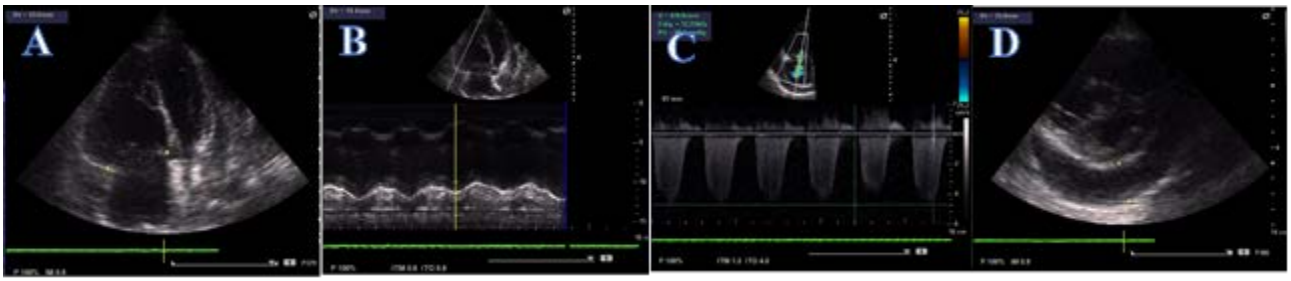

Figure 2. Trans thoracic cardiac Doppler ultrasound ((A) right ventricular dilation, (B) altered RV TAPSE, (C) presence of PAH, (D) profuse pericardial effusion).

because of the hemorrhagic risks, because the vessels are already weakened by the scleroderma lesion, in addition we do not have an adequate technical platform for the management of hemorrhagic complications.

All the clinical and paraclinical signs allowed us to make the diagnosis of cardiac involvement of scleroderma. Dermatologically, the scleroderma hypothesis was selected according to the 2013 classification criteria of the American College of Rheumatology (ACR) and the European League of Rheumatology for Systemic Scleroderma (EULAR).

Multi-disciplinary management was implemented, cardiologically it was put under diuretic (furosemide 40 mg/day), an IEC (enalapril 10 mg/day), salicylic acetyl acid 1000 mg 3 times/day, a phosphodiesterase inhibitor 5 (solagra 100 mg 1/4 tablet per day), a proton pump inhibitor (lanzocap 30 mg 1 tablet daily). Dermatologically she received: methotrexate 12.5 mg once a week (every Monday), folic acid 15 mg/week, an anti calcium (verapamil 240 mg 1 tablet per day), a corticosteroid (prednisone 30 mg 1 tablet daily).

After 5 months of treatment we note a favorable clinical course: remission of symptomatology including exercise dyspnea, retro sternal pain, digital ulcers have healed well, and paraclinical cardiac ultrasound shows a regression of pericardial effusion from 25 mm to 6 mm and hypertension from 80.4 mmHg to 33 mmHg.

3. Discussion

Cardiac involvement of systemic scleroderma can be primitive, related to systemic scleroderma itself or secondary to pulmonary arterial hypertension (PAH), pulmonary hypertension associated with infiltrating pneumonia causing dilation of the right cavities, or hypertension, most often accompanying renal involvement [7] [8]. Cardiac involvement occurs in 70% of cases during the first five years of the disease. Indeed, autopsy series show that nearly 80% of patients can have histological cardiac lesions [9] while cardiac involvement is most often silent and severe clinical translation is much less frequent. Thus, a so-called severe heart attack when there is heart failure, pericarditis or arrhythmia requiring treatment, would occur in only about 10% of patients with diffuse systemic scleroderma and about 1% of patients with limited systemic scleroderma [10]. If myocardial fibrosis is the main characteristic of the scleroderma heart, all cardiac structures can be affected, including the pericardium and more rarely the endocardium. Other manifestations are PAH and conduction or rhythm disorders, which are factors of very poor prognosis, sometimes causing sudden deaths [11].

The case we report is a young subject, whose symptomatologies began from the age of 19 years, marked by a retro sternal pain with type of gravity of progressive installation calmed by the leaning position forward or lying down, then dyspnea stage II of NYHA, puffiness of the face, edema of the lower limbs and cough. The physical examination had recovered a general condition preserved

with a performance index of WHO rated at 2, heart rate at 103 BPM, oxygen saturation: 98%. It presents a jugular turgor, the sign of Harzer in the xiphoid focus, on palpation the abdomen was distended with a painful hepatomegaly at 13 cm with smooth surface and foam edge, with a hepato jugular reflux, in the lower limbs, we note sock-shaped edema taking the bucket, on auscultation deafened BDC, a burst of B2 at the pulmonary focus, fine crackling gratings at the 2 pulmonary bases predominantly right. At the level of the mouth, it presents an ulceration of the lips. The particularity of our case, is its very young age compared to the majority of cases described by the literature, generally the cardiac involvement begins during the first 5 years of the disease [9]. Endothelial cells and their precursors, T and B lymphocytes, fibroblasts are involved in the onset of cardiac involvement through ischemic, inflammatory and fibroblast lesions of the myocardium, pericardium and conduction system [10]. Studies have shown that cardiac involvement is more frequent and may be histologically earlier than clinically, and even more so than ultrasound, as all patients in the study of Fernandes, *et al.* [12], which had normal cardiac ultrasound. Contrary to our case, cardiac involvement through clinical and ultrasound signs was the basis of the revelation of scleroderma.

Diastolic dysfunction of the left ventricle is one of the first ultrasound manifestations found by several authors, but this dysfunction was due to associated comorbidities and not to primary myocardial involvement. This diastolic alteration of the left ventricle was absent in our patient, which could be explained by the absence of comorbidity in our patient. Systolic dysfunction of the right ventricle, hypertension, pericardial involvements are other abnormalities frequently found on cardiac ultrasound, these signs were present in our patient. According to the authors this systolic dysfunction is usually secondary to scleroderma cardiomyopathy. The difference in impairment between the right and left ventricles is certainly partly related to the anatomical and physiological differences between the two ventricles. The right ventricle is thinner than the left ventricle and filling pressures are decreased. Thus, dilation of the right ventricle is probably earlier and easier to appear after volume overload, compared to the left ventricle. Systolic dysfunction of the right ventricle is a factor of poor prognosis. Anatomo-clinical studies show that conduction tissue is relatively spared by myocardial fibrose, and it is probably rather the diffuse myocardial fibrose that, in itself, inhibits cardiac conduction. Arrhythmias are also common during ScS and can be responsible for sudden deaths. Supraventricular tachycardia is the most common manifestation, while ventricular arrhythmia (extrasystoles or ventricular tachycardia) is rarer. It is due to myocardial fibrose, cardiac dysautonomia and damage to the conduction system. Kostis, *et al.* [13] reported that ventricular arrhythmias were present in 67% of patients assessed by an ECG holter. They were associated with a significant risk of sudden death and excess mortality. Our patient did not have an electrical abnormality on the ECG. On the therapeutic side, the patient was put under the treatment of heart failure (a conversion en-

zyme inhibitor, a beta blocker, a loop diuretic and an anti-aldosterone), salicylic acetyl acid 1000 mg 3 times/day, for pericardial effusion, a phosphodiesterase 5 inhibitor for the treatment of pulmonary hypertension, a proton pump inhibitor (lanzocap 30 mg 1 tablet per day) for gastric protection, a vasodilator (nifedipine after replaced by verapamil). Dermatologists introduced methotrexate 12.5 mg once a week (every Monday), folic acid and corticosteroid therapy (prednisone). Similar treatment has been reported in the literature for cardiac scleroderma [14]. The evolution was marked by a remission of symptomatology, a healing of digital ulcerations, and paraclinical regression of pericardial effusion and hypertension on cardiac Doppler ultrasound.

4. Conclusion

Cardiac involvement of ScS is not common in young subjects, its presence implies rapid, effective and multidisciplinary management to avoid further complications, which may compromise the life of the patient.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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