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Heart attacks in systemic scleroderma: about a case in Conakry

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Abstract

Summary: The objective of this study was to describe the cardiac manifestations during systemic scleroderma in Conakry and the therapeutic management.

Observation: 49-year-old female patient, admitted to the Medico-Surgical Emergency Department of Donka Hospital and University for chest pain, dry cough, osteo-articular pain, physical asthenia; evolution of symptoms for two weeks, no history of known cardiovascular disease and no cardiovascular risk factor. On physical examination, we have: TA=130/80 mmHg, heart rate at 110 beats/minute, height 1.78 cm, weight 65 kg (BMI: 20.56 kg/m²). There was also a decrease in heart sounds, no rhythm disorder or deformation of the chest, no scar on the thorax, temperature at 37.6°C, axial deformation of the fingers of both hands with hypochromia, hard, localized at the level of small joints.

Conclusion: Systemic scleroderma is an autoimmune disease that is more common in women than in men. His diagnosis is clinical, immunological and completed by the classification criteria. The functional or vital prognosis can be put into play especially in case of heart attack as was the case in our patient. Corticotherapy opens a window of hope despite the fact that treatment is difficult.

Keywords: Scleroderma, cardiac involvement, Conakry, young subject

Introduction

Systemic scleroderma is a rare autoimmune disease characterized by vascular involvement of the connective tissue (involvement of small vessels: arterioles and capillaries) responsible for systemic fibrosis that varies from person to person (digestive, cardiac, pulmonary, renal, articular involvement) [1].

The prevalence is estimated at about 1-9 / 100,000 for localized scleroderma and 1/6500 adults for systemic scleroderma, the disease is most prevalent in women, it usually occurs between 40 and 50 years of age and rarely affects children [1].

Cardiac involvement occurs in 70% of cases during the first five years of the disease [2].

Observation

49-year-old female patient admitted to the Medico-Surgical Emergency Department of the Donka Hospital and University for chest pain, dry cough, osteo-articular pain, physical asthenia; evolution of symptoms for two weeks, no history of known cardiovascular disease and no cardiovascular risk factor. On physical examination we noted: TA=130/80 mmHg heart rate at 110 beats/minute, height 1.78 cm, weight 65 kg (BMI: 20.56 kg/m²). There was also a decrease in heart sounds, no rhythm disorder or deformation of the chest, no scar on the thorax, the temperature at 37.6°C, there was axial deformation of the fingers of both hands with hypochromia, hard, localized at the level of small joints (**Figure 1**).

The electrocardiogram recorded a regular sinus rhythm at 68 cycles/min, and prior extended negative T waves with left ventricular electrical hypertrophy (**Figure 2**).

The frontal chest X-ray revealed an accentuation of the

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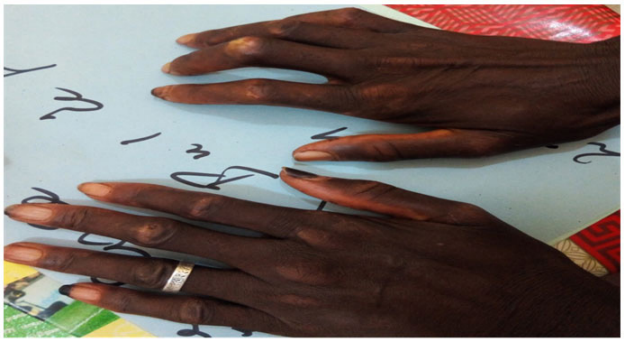

Figure 1. Axial deformation of the fingers with localized hardening of the skin of the hands.

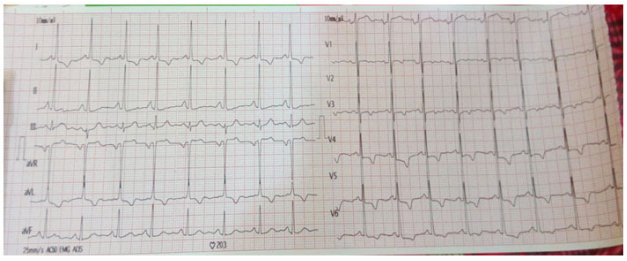

Figure 2. Regular sinus rhythm at 68 cycles/min, extended front T-waves extended with left ventricular hypertrophy.

broncho-vascular pattern, a diffuse heterogeneous heterameric opacity with bilateral hilar widening, and cardiomegaly with a cardiothoracic index of 0.69 (**Figure 3**).

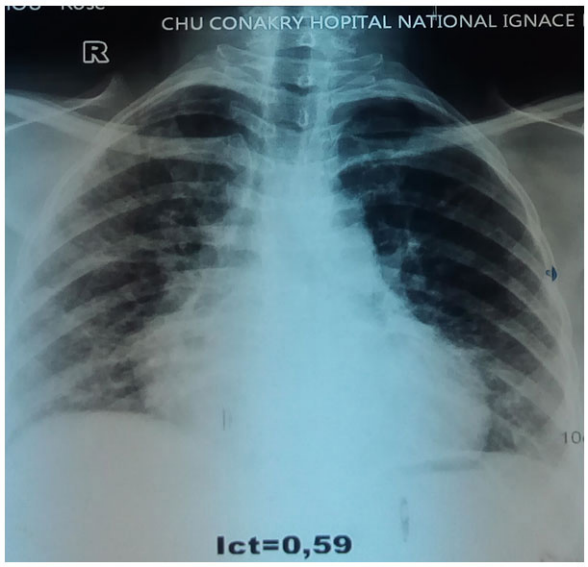

Figure 3. Digitized chest X-ray; highlights an accentuation of the bronchovascular framework, bilateral hilar widening, cardiomegaly with a cardiothoracic ratio at 0.69.

X-rays of the hands, wrists, and feet showed a pinch with osteo-condensations of inter-carpal, inter-tarsal, carpo-metacarpal, tarso-metatarsal, metacarpophalangeal, metatarsophalangeal, and inter-phalangeal joint space advocating for arthritis (**Figure 4**).

Cardiac Doppler ultrasound revealed a circumferential circumferential pericardial effusion of moderate to moderate abundance (11mm to 18mm) in anteroposterior without fibrin network (**Figure 5**).

Biology: Sclero-70 greater than 9U (normal value less than

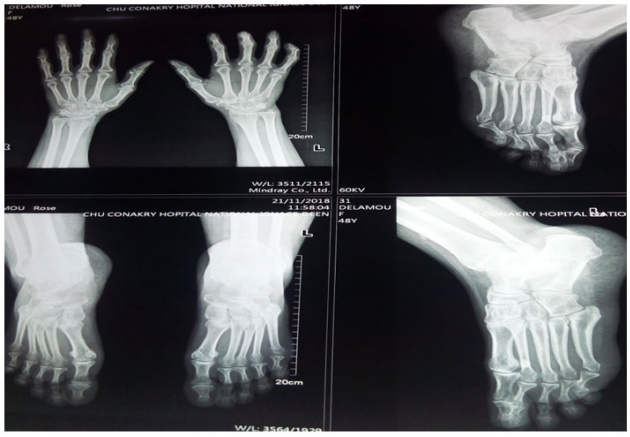

Figure 4. X-ray of the hands and wrist plus feet showing a pinch.

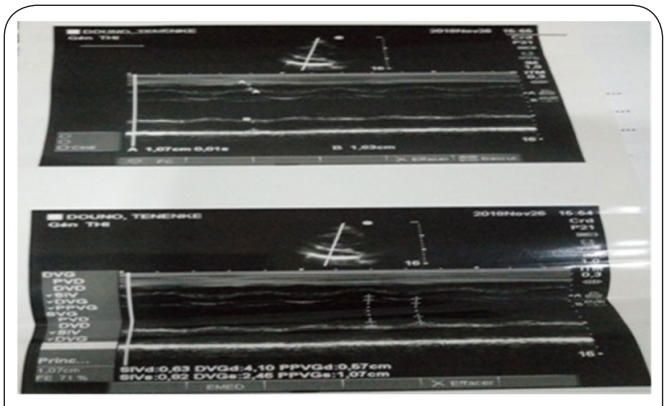

Figure 5. Circumferential fluid pericardial effusion of moderate to moderate abundance of 11 to 18 mm in anteroposterior without fibrin network.

0.9U), rheumatoid factor was negative, Hemoglobin=13.3 g/dl, neutrophils=49.7%, eosinophilic polynuclear=3.5%, polymorphonuclear basophilic=0.9%, hyper lymphocytosis=41.1%, hyper monocytosis=4.8%, platelets=191 g/l, blood Ionogram (Natrium=5.9 mmol/l, potassium=129 mml/l, calcium=2.36 mmol/l, Cl-=98 mmol/l HCO-3=26 mmol/l.

Discussion

Several works have been devoted to the cardiac lesions of

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scleroderma whose main mechanism is a disruption of the functioning of the cardiac muscle by a bad vascular irrigation. It remains for a long time without or with few clinical symptoms that may cause shortness of breath and/or chest pain, see pericarditis [3].

We report the case of a 49-year-old woman who developed systemic scleroderma complicated by anteroposterior pericardial effusion. This condition mainly affects women (4 women to 1 man approximately), and usually occurs between 40 and 50 years. Scleroderma can occur exceptionally in children. It affects all populations in principle, but is more common in people exposed to certain industrial chemicals or in miners [4]. In case of scleroderma, collagen is produced in excess and accumulates in the skin and various organs. This is called fibrosis, in reference to the fibrous and rigid aspect that gives excess collagen to the affected organs, including the skin. Fibrosis can also thicken the walls of some vessels to the point where it eventually clogs them and prevents blood from circulating. In particular, these lesions threaten the internal organs (heart, kidneys) by partially depriving them of oxygen and the nutrients normally provided by the blood, and a rare disease [4].

Our case is a perfect illustration of this nosological rarity. Predisposing factors are found in the majority of cases, including certain industrial chemicals or juveniles, and gender [5].

In our case, pericardial effusion was clinically manifested by chest pain, cough, fever, physical asthenia, and bone manifestation was osteoarticular pain and axial deformity of the fingers. Indeed, autoptic series show that nearly 80% of patients may have histological cardiac lesions [3]. Pericardial involvement is very common in autoptic studies in 33 to 72% of patients, whereas it is clinically evident in only 11 to 41%. Tamponade is exceptional. In cardiac ultrasonography, the prevalence of pericardial effusion varies between 11 and 41%, but most recent studies give a figure between 11 and 18%. Most often, these are fortuitous ultrasound findings, without clinical translation [1]. Our clinical case is an example of this as shown (**Figure 3**): Circumferential fluidic pericardial effusion of moderate to moderate abundance (11mm to 18mm) in anteroposterior without fibrin network. Normal sized heart chambers free of thrombus. Good bi-ventricular systolic function FE=71% in Teicholz, TAPSE =18mm. In our case the treatment was: administration of calcium + vit D 1 g/day and deparasitant based on albendazol 400 mg per day for three days, Colchicine 20 mg at the dose of 60 mg per day and divided in three days, the patient benefited from cortisone (diprostene) infiltration at four joints (two wrists and two ankles). After one week of treatment there was a clear remission of signs that eventually disappeared after 6 months of evolution.

Conclusion

Systemic scleroderma is a more common connective tissue disease in women than in men.

His diagnosis is clinical, immunological and completed by the classification criteria.

The functional or vital prognosis can be put into play especially in cases of cardiac damage as was the case in our patient. Corticotherapy opens a window of hope despite the fact that treatment is difficult.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

Authors' contributions	DS	AK	ISS	AS	AC	ISB	IC	BFD	MB	MDB
Research concept and design	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Collection and/or assembly of data	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Data analysis and interpretation	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Writing the article	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Critical revision of the article	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Final approval of article	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Statistical analysis	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓

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