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**CREST syndrome. Report of a Clinical Case**

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**ABSTRACT**

**Introduction:** CREST is a rheumatic inflammatory disease that constitutes a clinical form of systemic scleroderma manifested by Calcinosis, Raynaud's phenomenon, Esophageal dysmotility, Sclerodactyly, Telangiectasia.

**The Case:** 25-year-old female patient, white skin color; with a history of hypothyroidism who attended the Internal Medicine service for presenting a painful joint condition that affected both wrists, small joints of the hands (metacarpophalangeal and proximal interphalangeal), morning stiffness for more than 1 hour. Raynaud's phenomenon for approximately 2 years, difficulty to eat solid food, epigastralgia that is exacerbated by eating food. An X-ray of hands is performed showing calcinosis at the level of the distal phalanges.

**Conclusion:** This is an interesting and peculiar case in which we found an incomplete CREST syndrome in which the patient evolved satisfactorily with the indicated treatment.

**Key words:** Systemic Scleroderma, CREST Syndrome.

**RESUMEN**

**Introducción:** CREST es una enfermedad inflamatoria reumática que constituye na forma clínica de esclerodermia sistémica que se manifiesta por: Calcinosis, Fenómeno de Raynaud, Dismotilidad esofágica, Esclerodactilia, Telangiectasia.

**Reporte de caso:** Paciente femenina de 25 años, color de piel blanca; con antecedentes de hipotiroidismo que acude al servicio de Medicina Interna por presentar un cuadro doloroso articular que interesaba ambos carpos, pequeñas articulaciones de las manos (metacarpofalángicas e interfalángicas proximales), rigidez matutina mayor de 1 hora, fenómeno de Raynaud desde aproximadamente 2 años, dificultad para ingerir alimentos sólidos, epigastralgia que se exacerba al ingerir alimentos. Se realiza una Radiografía de manos que muestran calcinosis a nivel de falanges distales.

**Conclusión:** Se trata de un caso interesante y peculiar en el que encontramos un síndrome de CREST incompleto en el que la paciente evolucionó de manera satisfactoria ante el tratamiento indicado.

**Palabras clave:** Esclerodermia Sistémica, Síndrome CREST.

**INTRODUCTION**

Systemic sclerosis (SS) is a multisystem disease characterized by widespread vascular dysfunction and progressive fibrosis of the skin and internal organs. It is a rare and chronic rheumatic pathology that mainly affects females between the fourth and fifth decade of life 1.

Genetic, infectious and environmental factors play a key role; vascular injury, fibrosis and immune activation are the main pathogenic factors. The global incidence is 0.6 to 122 cases per million people per year. There are differences in incidence depending on region, sex, and race: it is higher in the US and Australia than in Japan or Europe; between three women for each man with this disease; greater in the black race than in the white 2.

Scleroderma is usually classified into two large groups: diffuse scleroderma, where it is common to find skin sclerosis at the proximal and distal level of the extremities, face, and trunk, as well as pulmonary, renal, gastrointestinal, and cardiac involvement. The other group is limited scleroderma, which has a better prognosis than diffuse scleroderma, since organ and system involvement is to a lesser degree and intensity 3.

The pathogenesis of systemic sclerosis is not entirely clear, but it is believed that the clinical manifestations are the product of alterations in the functionality of certain cells such as fibroblasts, endothelial cells, and T and B lymphocytes, which end up causing cutaneous and visceral fibrosis. , damage to the microvasculature and alterations in humoral immunity, with the consequent production of autoantibodies, of the anti-SCL70 and anti-centromere type 2,4.

Patients with limited sclerosis, also known as CREST (Calcinosis, Raynaud's phenomenon, Esophageal Dysmotility, Sclerodactyly, Telangiectasia) usually present with Raynaud's phenomenon long before developing clinical symptoms of the disease. Currently, only three of the criteria are necessary to make the diagnosis (incomplete CREST). They also tend to develop fibrosis of the skin, generally circumscribed to the fingers and regions distal to the elbows and knees, pulmonary fibrosis is usually moderate, pulmonary hypertension is usually late-onset; while renal involvement is extremely rare, and the characteristic autoantibodies are usually anticentromere 5,6.

**Scientific problem**

Clinical and radiological findings that allowed the diagnosis of CREST Syndrome in a patient from HGD Comandante Pinares.

**Justification**

Taking into account that it is not a frequent disease, it was decided to carry out this report to describe the clinical picture, the complementary tests, the treatment of this patient and to share these findings with the medical community in general. The recognition of CREST syndrome as a distinct diagnostic entity is necessary not only for nosological classification but also because it leads to a change in the way the patient is approached and managed.

**OBJECTIVES**

**General objective:**

Describe the clinical characteristics that allowed establishing the diagnosis and treatment of CREST Syndrome in a patient.

**Specific objectives:**

1. Identify the clinical manifestations of said nosological entity.
2. Describe the diagnosis and treatment used in the clinical case.

**CASE PRESENTATION**

A 25-year-old female patient, white skin color; with a history of Hypothyroidism who attended the Internal Medicine service of the Comandante Pinares General Hospital in June 2021 due to a painful joint condition that affected both wrists, small joints of the hands (metacarpophalangeal and proximal interphalangeal joints), morning stiffness for more than 1 hour. Raynaud's phenomenon, difficulty eating solid food, epigastric pain that is exacerbated by eating food, and intermittent unquantified fever of the same duration, with chills and profuse diaphoresis.

On physical examination: presence of hyperchromic lesions on the back of both hands (See Figure 1), livedo reticularis, accompanied by induration of the skin in the palmar region of both hands, sclerodactyly, there was difficulty pinching the skin; Raynaud's syndrome (approximately 2 years of evolution). In the lower extremities, rough-looking lesions are also observed on the dorsum of both feet. An X-ray of the hands showing calcinosis at the level of the distal phalanges (See Figure 2) is performed, a chest X-ray is also performed without yielding any pathological findings.

**Table 1.** Laboratory studies.

|  |  |
| --- | --- |
| Complementary | Results |
| Hemoglobin | 13.0 g/L |
| Hematocrit | 0.37 |
| Erythrocyte sedimentation rate | 58 mm/h |
| Platelet Count | 220x109/L |
| Leukogram | 7.8x109/L |
| TGP | 25 U/L |
| TGO | 34 U/L |
| Cholesterol | 3.4 mmol/L |
| Triacylglycerides | 1.6 mmol/L |
| Serology | NR |
| PCR | 1.3 mg/dL |
| Rheumatoid factoy | 80 UI/mL |
| IgG | 24.3 g/L |
| IgA | 7.1 g/L |
| IGM | 3.3 g/L |
| C3 | 1.7 g/L |
| C4 | 0.3 g/L |
| Peripheral Sheet: | Normochromic, normocytic,  leukocytes and platelets  suitable |

Source: Cynical History

It was decided as a treatment for the patient the use of gloves on the hands and feet; Avoid stress and smoking. Artificial tears 1 drop every 4 hours. Calcium channel blockers: Nifedipine 10m every 12 hours (prior BP control); lubricating creams for the skin, Vitamin E 1 tablet per day and Colchicine 1 tablet per day; noting clinical improvement in the patient and evolving satisfactorily.

**DISCUSSION**

The classification criteria for systemic sclerosis were developed by a joint committee of the American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR) to identify patients with SS in 2013 7. These criteria showed more sensitivity (0 .91) and specificity (0.92) than the guidelines previously carried out 7.8.

According to ACR-EULAR, thickening of the skin of the fingers of both hands with extension to the metacarpophalangeal joints is a sufficient criterion for diagnosis 8. Sclerodactyly is due to excessive fibrosis that generates constant flexion of the fingers of the hand and can be present in other pathologies such as diabetes mellitus 1; our patient presented difficulty in extending the fingers of the hands and did not present any criteria for DM1 9 .

Raynaud's Syndrome is vasospasm, induced by cold and stress, of the digital arteries and cutaneous arterioles; It can be present in healthy people. After the appearance of this phenomenon, patients may otherwise be asymptomatic for years, as in the case presented, or other symptoms and signs of the disease may rapidly develop 2,5. Other microvascular manifestations that we can see in SS are lesions on the tips of the fingers, telangiectasias, capillaroscopy abnormalities, erectile dysfunction, and kidney disease 8,10.

Nearly 90% of patients with SS have some degree of gastrointestinal involvement and approximately half are symptomatic 11. Although the esophagus is the most affected part of the gastrointestinal tract, as is the case under study, any part of the gastrointestinal tract can be involved. The presence of malabsorption and esophageal dysfunction among patients with SS is associated with an unfavorable prognosis 12.

Between 25 and 40% of patients will develop calcinosis cutis ten years after the onset of the disease. This is produced by a dystrophic calcification with an accumulation of hydroxyapatite crystals at the level of the subcutaneous tissue located in areas of friction, among the most common we find those present in the case: knuckles of metacarpophalangeal joints, back of hands13.

In general, treatment is symptomatic depending on which organ is involved. For example, patients with Raynaud's phenomenon are treated with calcium channel blockers, patients with renovascular hypertension are treated with angiotensin converting enzyme inhibitors, etc. 14. The patient had an evident clinical and symptomatic improvement with the therapeutic management provided. and progressed satisfactorily.

**CONCLUSIONS**

In this case, the main clinical findings that allowed us to focus the diagnosis on this disease were sclerodactyly, Raynaud's syndrome, calcinosis, and esophageal dysfunction; those that constitute 4 of the 5 characteristics that make up the Syndrome (an incomplete CREST). The evaluation included X-rays of the hands and wrists, and laboratory tests that indicated hibergammaglobulinemia and positive RF. This disease continues to be incurable, so treatment is based on acute symptom control, medical prevention of complications, and rehabilitation. The patient evolved satisfactorily after the indicated treatment.

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**ANNEXES**

 Figure 1: Presence of lesions on the back of both hands: livedo reticularis, accompanied by induration of the skin in the palmar region of both hands, sclerodactyly, and Raynaud's syndrome.



Figure 2: X-ray of hands showing calcinosis at the level of the distal phalanges.

