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Scleromyositis: about a case of early diagnosis in Lomé and review of the literature

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ABSTRACT

Introduction: Scleromyositis (SM) represents an overlap syndrome between scleroderma and inflammatory myopathy. It is the most common of the overlapping syndromes.

Observation: The authors report a case of scleromyositis in a 26-year-old woman. The diagnosis was facilitated by the presence of myalgia in the anterior aspect of the left leg, as detected by magnetic resonance imaging (MRI) of the left thigh, which revealed edema involving the intermedial vastus and the medium gluteal muscles. Muscle enzymes were normal, but electroneuromyogram revealed a neuropathic pattern. Furthermore, antinuclear antibodies (ANA) were found to be positive using indirect immunofluorescence on Hep 10-20 slides, with a titer of 1/160, a speckled appearance and anti-polymyositis/scleroderma antibodies were identified. The patient was treated with corticosteroids and then immunosuppressants, with a favorable clinical response.

Conclusion: Muscle MRI can facilitate the diagnosis of scleromyositis in countries where access to muscle biopsy is challenging.

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INTRODUCTION

Scleromyositis (SM) is an overlap syndrome involving two distinct autoimmune diseases: systemic sclerosis and either polymyositis (PM) or dermatomyositis (DM) [1]. It is the most common of the overlap syndromes [2]. Autoimmune diseases in sub-Saharan Africa are characterized by delayed diagnosis [3]. In Togo, a 10-year study conducted in Lomé confirmed the rarity of scleroderma, which was the second most common connective tissue disease [4]. We felt it appropriate to report a rare case of scleromyositis that was diagnosed early.

OBSERVATION

This is a young patient, 26 years old, right-handed, of black race, residing in Lomé, who consulted in August 2023 for left crural pain.

Symptoms began in October 2022 with left thigh muscle pain after skipping. Two months later, the pain intensified and became inflammatory. Difficulty rising from a squatting to a standing position also developed. Her personal and family medical history is minimal, with no history of consanguinity among her parents.

A consultation was conducted at a health center in Lomé, where a standard hip X-ray was performed, which was normal, as were the blood tests (complete blood count, erythrocyte sedimentation rate, and creatine phosphokinase). She was treated with

paracetamol, resulting in a transient improvement in pain. A muscle MRI of the thighs performed in June 2023 revealed a focal T2 and STIR hyperintense area (edema) involving the left vastus intermedius and left gluteus medius muscles at the level of the greater trochanter (Figure 1).

The patient was referred to a neurology consultation and the examination revealed a weight of 55 kg and a height of 154 cm, resulting in a body mass index (BMI) of 23.2. The neurological examination was entirely normal. Examinations of the skin, digestive system, and other organs were also normal. The clinical picture progressed with constipation in September 2023 and gastroduodenal reflux (GERD) in November 2023. She did not report any Raynaud's phenomenon. The diagnostic hypotheses were inflammatory, infectious, or hereditary myopathy.

The blood tests in August 2023 showed normal creatine phosphokinase (CPK), lactate dehydrogenase, and myoglobin levels; a normal complete blood count; a normal erythrocyte sedimentation rate (ESR) of 20 mm/hr; normal blood glucose, creatinine clearance, and liver function tests; and a normal serum protein electrophoresis profile. The electrocardiogram was normal. The electroneuromyogram (ENMG) revealed a multiple mononeuropathy with neurogenic patterns in some areas (Figures 2 to 5).

The etiological assessment of this myopathy included an indirect immunofluorescence assay of antinuclear antibodies which were positive with a titer of 1/160, a speckled appearance, negative anti-DNA autoantibodies, negative anti-soluble nuclear antigen (ENA) autoantibodies (search carried out simultaneously for anti-Soluble Nuclear Ribonucleoprotein A (anti-SSA/Ro), anti-Sjögren syndrome B (SSB/La), anti-Smith (SmD1), Uracil 1 Small nuclear ribonuclear protein (U1snRNP), anti-aminoacyl-t-RNA-synthetases (Jo1), anti-topoisomerase I (Scl 70) and centromere.

The assay of specific autoantibodies for myositis was weakly positive for anti-polymyositis/scleroderma antibodies (anti-PM/Scl), negative for anti-threonyl-tRNA synthetase (PL7), alanyl-tRNA synthetase (PL12), glycine-tRNA synthetase (EJ) isoleucyl-tRNA synthetase (QJ), Signal Recognition Particle (SRP), Nucleosome remodeling histone deacetylase complex (Mi2), Ku, Transcriptional intermediary factor 1 gamma (TIF1 gamma), anti-Melanoma differentiation-associated gene 5 (MDA5), NXP2 Nuclear matrix protein-2 (NXP-2), Small ubiquitin-like modifier activating enzyme (SAE).

The chest CT scan requested to look for diffuse interstitial lung disease came back normal.

A diagnosis of scleromyositis was made, and the patient was treated with oral prednisone 60 mg in the morning for one month, followed by a taper of 10 mg per week, discontinued after six weeks. Oral methotrexate 7.5 mg/week was introduced after the first month. She also received omeprazole 20 mg in the morning for three months. At three months, the patient showed improvement in muscle pain and GERD.

DISCUSSION

Historically, scleromyositis has been defined as an overlap syndrome (OS) between scleroderma and inflammatory myopathy (IM) [1], [2]. For some authors, SM is a disease that involves more than just an overlap syndrome between autoimmune myositis and systemic scleroderma, and they considered it important to highlight the clinical, serological, and histopathological characteristics of SM [5]. Scleroderma is the most frequent of the overlap syndromes, representing 42.6% of IM cases in the series by Troyanov et al [2] . These results are consistent with those of African studies. Thus, in Senegal, scleroderma represented 50% of OS cases [6], while in Durban, South Africa, scleroderma accounted for 63.4% of OS cases [7].

Myalgia was the reason for the patient's consultation. Extramuscular symptoms, which were solely digestive in nature, appeared much later. Myalgia is found in 21 to 88% of patients with SM [5]. In an American study, the appearance of muscle weakness was demonstrated in the 6-year follow-up (37% at the beginning, 93% at the end) [8]. Muscle weakness was more frequent in the arm abductors and hip flexors and can develop during the course of the disease [8].

Scleromyositis is distinguished from scleroderma without muscle involvement by the frequency of involvement of other organs: such as the lungs, leading to diffuse interstitial lung disease (up to 68% in SM versus 13% in scleroderma), myocardial involvement (up to 21% versus 10%), scleroderma renal crisis (15% versus 5%), more diffuse than limited skin thickening (40-75% versus 25-60%, respectively), tendon crepitus and synovitis [5].

Gastrointestinal manifestations of scleroderma include dysphagia, acid reflux, gastric ulcers, retrosternal pain, and constipation [9] . However, gastrointestinal manifestations are often more severe in patients with scleromyositis compared to patients without myositis [5].

Paraclinical findings show elevated creatine phosphokinase (CPK) levels in 8% to 100% of patients with scleromyositis, while aldolase levels are elevated in 11% to 75% of these patients [5]. Electroneuromyography is abnormal in 11% to 92% of scleromyositis cases. The myopathic pattern is the most frequently observed, predominantly affecting proximal rather than distal muscles. The neuropathic profile has also been reported in 18 to 29% of patients with scleromyositis. In our patient, CPK and aldolase levels were normal, but the ENMG revealed a neuropathic profile.

Muscle MRI was the diagnostic tool, revealing edema of the left gluteus medius and vastus intermedius. MRI allows noninvasive visualization of the characteristic changes of myositis (edema, fat replacement, atrophy, fasciitis) [10] . In patients with SM, myositis lesions on MRI include fasciitis (thickening and/or increased signal intensity of the fascia on STIR and post-gadolinium images) and/or perifascial edema and/or muscle edema [5].

Muscle biopsy can reveal lesions specific to scleromyositis, such as vasculopathy with significant reduction of the basement membrane. It can also reveal serious features such as muscle fiber necrosis or fibrosis [5]. In countries where muscle biopsy is difficult to access, such as ours, muscle MRI could help guide the diagnosis.

Historically, the anti-PM-Scl antibody was considered the serological marker for SM. The PM/Scl antigen is located at the ribosome assembly site in the nucleolus. Although its function is not fully understood, the PM/Scl complex may play a role in ribosome maturation. Autoantibodies are directed primarily against two molecules of 100 kDa and 75 kDa [11] . Currently, the prevalence of anti-PM/Scl autoantibodies in SM is 31% [5] . Other autoantibodies associated with MS include anti-U1RNP (10%–46%), anti-Ku (38%–55%), anti-U3RNP, and anti-RuvBL1/2, which are less common. It is worth noting that seronegative forms exist in 50% of SM cases [5]. The unusual aspect of this observation lies in the relatively early diagnosis in a geographical area where autoimmune diseases are often characterized by delayed diagnosis [3] . In Senegal, the average diagnostic delay was 38.6 months [3]. In our case, the diagnosis of myositis was established at a stage of myalgia, in the absence of elevated muscle enzymes and well before the onset of digestive symptoms. Furthermore, it is interesting to note that if we apply the American Rheumatology Association (ACR) diagnostic criteria for scleroderma to our patient, the score is 0 [12]. Therefore, the specific diagnostic criteria for scleromyositis warrant codification.

Corticosteroids, cyclophosphamide, mycophenolate mofetil, tocilizumab, rituximab, nintedanib, and hematopoietic stem cell transplantation have demonstrated therapeutic efficacy in patients with systemic sclerosis [5]. However, there are no consensus guidelines for the management of scleromyositis. In the context of MS, the benefits of corticosteroids must be weighed, as corticosteroids have been associated with an increased risk of renal crisis in scleroderma. Consistent with their association with necrosis and/or inflammation on muscle biopsy, anti-PM/Scl, -U1-RNP, and -Ku autoantibodies have been associated with a good response of myositis to corticosteroids [5]. Methotrexate and azathioprine have also been used. Efficacy and corticosteroid-sparing effect have only been demonstrated for methotrexate in the context of juvenile dermatomyositis [13].

The patient was treated orally, initially with 60 mg of Prednisone for 1 month, then Methotrexate 7.5 mg per week with a favorable evolution on the myalgias and digestive symptoms.

It should be noted that this observation has limitations, namely the lack of data on the patient's long-term follow-up and the potential generalizability of scleroderma. Prognostically, it has been shown that patients with SM have a lower survival rate than patients with scleroderma without myositis, with cardiopulmonary disease being the most frequent cause of death (42% to 63%) [5]. Furthermore, in patients with scleromyositis, complications related to scleroderma accounted for up to half of the deaths.

CONCLUSION

Scleromyositis, or the overlap syndrome between systemic scleroderma and inflammatory myopathy, is a recently recognized entity whose diagnostic criteria warrant codification. Myalgia in a young woman can be the presenting symptom. Muscle MRI can guide the diagnosis in countries where access to muscle biopsy is limited.

ICONOGRAPHY

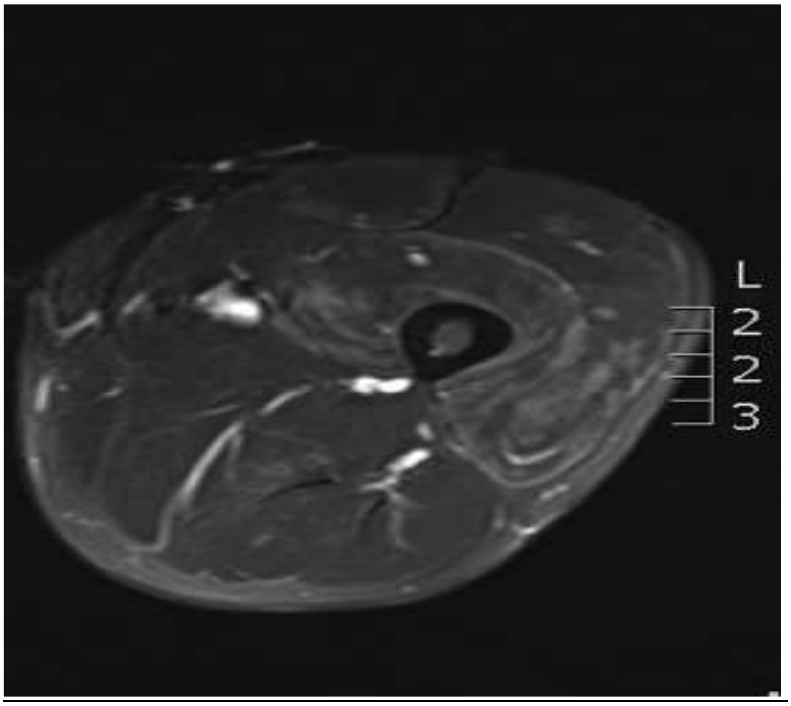

Figure 1: Axial T2-weighted MRI of the left thigh muscle showing a hyperintense signal in the left vastus intermedius representing edema.

Motor nerve conduction			
Nerves	Distal latency (ms)	Amplitude (mV)	Velocity (m/s)
Right median	4.7	2.8	54.1
Left median	2.5	1.9	54.2
Right ulnar	3.2	6.7	81.0
Left ulnar	2.7	6.4	74.4
Right common fibular	3.4	4.1	52.5
Left common fibular	3.4	3.8	55.0
Right tibial	5.2	6.1	49.2
Left tibial	5.6	3.2	40.7

Figure 2: Electroneuromyographic examination: motor conduction (examination showing collapsed motor amplitudes in the left common fibular left median and rigth median nerves)

Sensitive nerve conduction (orthodromic technique)			
Nerves	Distal latency (ms)	Amplitude (µV)	Velocity (m/s)
Right radial	2.3	47.1	60.9
Left radial	2.0	48.6	70.0
Right median	1.6	44.2	68.8
Left median	2.0	41.7	50.0
Right ulnar	1.5	30	60.5
Left ulnar	1.7	32.8	52.9
Right sural	2.6	45.9	46.2
Left sural	2.3	62.4	52.2

Figure 3: Electroneuromyographic examination : sensory conduction (examination within normal limits)

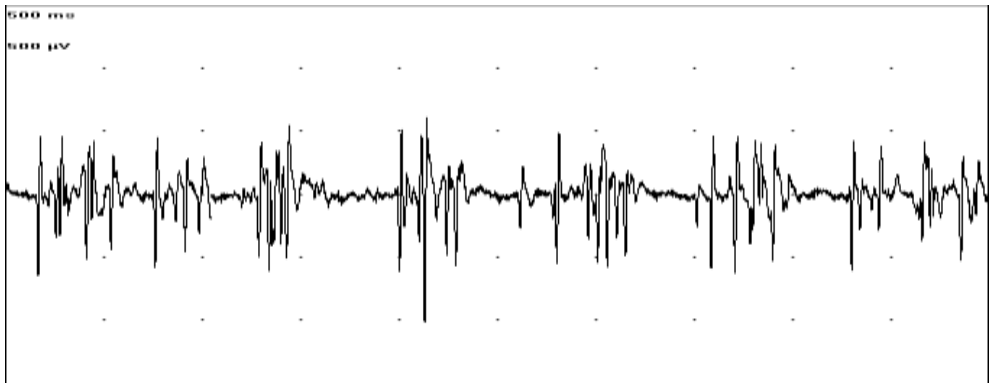

Figure 4: Neurogenic tracing of the right vastus lateralis (maximum effort)

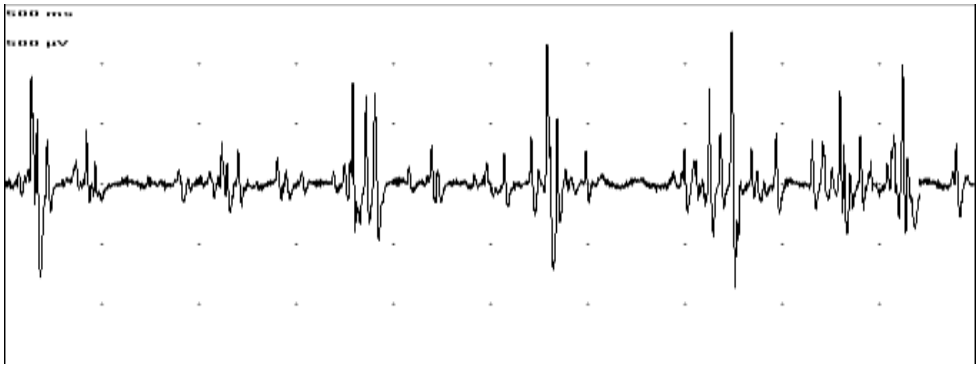

Figure 5: Neurogenic tracing of the left vastus lateralis (maximum effort)

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Conflict of interest
There is no conflict of interest

Consent

Consent has been taken

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