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Bulbar Syndrome Revealing Anti-PM/SCL Antibody-Positive Scleromyositis: A Case Report

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Abstract

Scleromyositis is a new clinical entity, an overlapping syndrome involving two distinct autoimmune diseases, systemic scleroderma and polymyositis or dermatomyositis. We report an observation of a 43-year-old female patient with a history of thyroidectomy, who has been presenting with phonation and swallowing disorders for over 6 months. Clinical examination revealed neuro-myogenic syndrome. Biological findings included elevated creatine phosphokinase at 1497 IU/L and lactate dehydrogenase at 954 IU/L, and the presence of a chronic inflammatory syndrome on serum protein electrophoresis. The electroneuromyography showed sensitive and motor polyneuropathy associated with polymyositis. Immunological tests revealed positive anti-PM/Scl-75 antibodies in the DOT myositis assay. The patient was treated with corticosteroid therapy 1mg/kg/day plus adjuvant plus methotrexate 15mg/week. The evolution was marked by a progressive clinical improvement of the motor deficit and biological improvement with a clear decrease in creatine phosphokinase.

Antibodies anti-PM75-scl | Diaphragmatic dysfunction | scleromyositis

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Abstract

Scleromyositis is a new clinical entity, an overlapping syndrome involving two distinct autoimmune diseases, systemic scleroderma and polymyositis or dermatomyositis. We report an observation of a 43-year-old female patient with a history of thyroidectomy, who has been presenting with phonation and swallowing disorders for over 6 months. Clinical examination revealed neuromyogenic syndrome. Biological findings included elevated creatine phosphokinase at 1497 IU/L and lactate dehydrogenase at 954 IU/L, and the presence of a chronic inflammatory syndrome on serum protein electrophoresis. The electroneuromyography showed sensitive and motor polyneuropathy associated with polymyositis. Immunological tests revealed positive anti-PM/Scl-75 antibodies in the DOT myositis assay. The patient was treated with corticosteroid therapy 1mg/kg/day plus adjuvant plus methotrexate 15mg/week. The evolution was marked by a progressive clinical improvement of the motor deficit and biological improvement with a clear decrease in creatine phosphokinase.

Keywords: *Antibodies anti-PM75-scl, Diaphragmatic dysfunction, scleromyositis*

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1. Introduction

Scleromyositis is an overlap syndrome combining two distinct entities: systemic sclerosis and either polymyositis or dermatomyositis [2, 5]. It is a rare autoimmune disease characterized by relapses and remissions [4, 9]. It may be life-threatening due to bulbar, respiratory, and/or cardiac involvement [4, 9]. We report a case managed in our medical neuro-intensive care unit involving a patient with scleromyositis and positive anti-PM75 antibodies. The outcome was favorable with corticosteroid therapy and methotrexate.

2. Clinical Observation

This was a 43-year-old woman, in a monogamous marriage, mother of six children, and a housewife. Her medical history included a subtotal thyroidectomy performed in October 2021, followed by treatment with Levothyrox 200 μg/day.

Nine months before admission, she developed phonation disorders characterized by dysphonia associated with swallowing difficulties, which developed gradually and progressively worsened without fluctuation over time. Four months later, she progressively developed motor weakness affecting all four limbs, predominantly involving the proximal muscles.

The clinical picture evolved in a context of inflammatory polyarthralgia involving the peripheral joints (knees, elbows, ankles, and hands), associated with myalgia.

Physical examination on admission revealed a neuromyogenic syndrome involving all four limbs, with proximal-predominant motor weakness, more marked in the upper limbs; thenar and hypothenar muscle wasting; limited mouth opening; a pseudobulbar syndrome; dropped head syndrome; bilateral pitting edema of the lower limbs; reduced chest expansion; and absent idiomuscular reflexes at the quadriceps.

On extra-neurological examination, the patient presented with erythematous lesions on the eyelids and symmetrical erythematous scaly lesions over the metacarpophalangeal and interphalangeal joints.

After five days of hospitalization, the clinical course was marked by the development of acute respiratory distress requiring transfer to the neurological intensive care unit.

Laboratory investigations revealed elevated creatine phosphokinase (CPK) at 1,271 IU/L and lactate dehydrogenase (LDH) at 954 IU/L (Figure 1), evidence of a chronic inflammatory syndrome on serum protein electrophoresis, and elevated C-reactive protein.

Renal function, thyroid hormone levels, serum calcium, and serum albumin were normal. Arterial blood gases showed mild hypercapnia at 50 mmHg and hypoxemia at 60 mmHg.

The immunological work-up revealed positive anti-PM75 antibodies on the myositis DOT assay (Figure 2).

A cervico-thoraco-abdomino-pelvic CT scan (Figure 3) showed micronodules and nodules of varying sizes in the basal regions of the lungs, associated with a 10-mm right cervical lymph node,

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RESULTATS

EXAMEN	RESULTAT	NORME
LDH	545	190 - 415 U/L
CPK	356	20 - 170 U/L
ASAT (TGO)	94	< 40 UI/L
ALAT (TGP)	112	< 41UI/L

Figure 1. Initial CPK and LDH results

IMMUNOLOGIE
Auto–Immunité

Marqueurs des myosites (Dot–Euroimmun–BioAdvance) #

Anticorps anti–Mi2 alpha	**Négatif**
Anticorps anti–Mi2 beta	**Négatif**
Anticorps anti–TIF1 gamma	**Négatif**
Anticorps anti–MDA5	**Négatif**
Anticorps anti–NXP2	**Négatif**
Anticorps anti–SAE	**Négatif**
Anticorps anti–Ku	**Négatif**
Anticorps anti–PM 100	**Négatif**
Anticorps anti–PM 75	**Positif**

Faible réactivité. Résultat à confronter au contexte clinique ainsi qu'à l'ensemble du bilan auto–immun (ASPECT DE FLUORESCENCE SUR HEP2/2000 notamment).
Réactivité non spécifique PROBABLE.

Anticorps anti–Jo1	**Négatif**
Anticorps anti–SRP	**Négatif**
Anticorps anti–PL7	**Négatif**
Anticorps anti–PL12	**Négatif**
Anticorps anti–EJ	**Négatif**
Anticorps anti–OJ	**Négatif**

Réalisé par Biomnis IVRY – **Validé par :** Dr Laurence GUIS–CABANIE

Figure 2. Myositis DOT assay results

suggestive of interstitial lung disease. Echocardiography was normal.

Electroneuromyography (Figures 4 and 5) showed an axonal sensorimotor polyneuropathy associated with myogenic patterns, particularly involving proximal myotomes, suggesting polymyositis.

The diagnosis of scleromyositis with positive anti-PM75 antibodies was therefore established.

The patient was treated with corticosteroid therapy at 1 mg/kg/day with adjunctive treatment, together with methotrexate at 15 mg/week. The clinical course was characterized by progressive improvement in motor weakness and laboratory parameters, with a decrease in CPK and LDH levels (Figure 6). The modified Rankin Scale score at 6 months was 0.

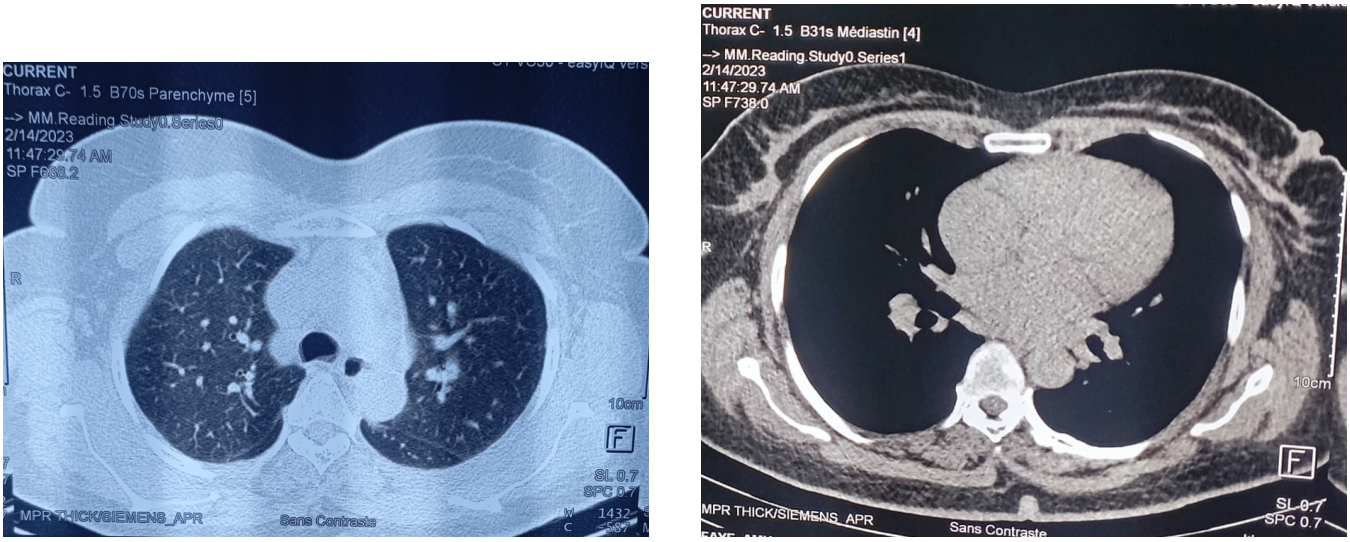

Figure 3. Chest CT angiography showing interstitial lung disease

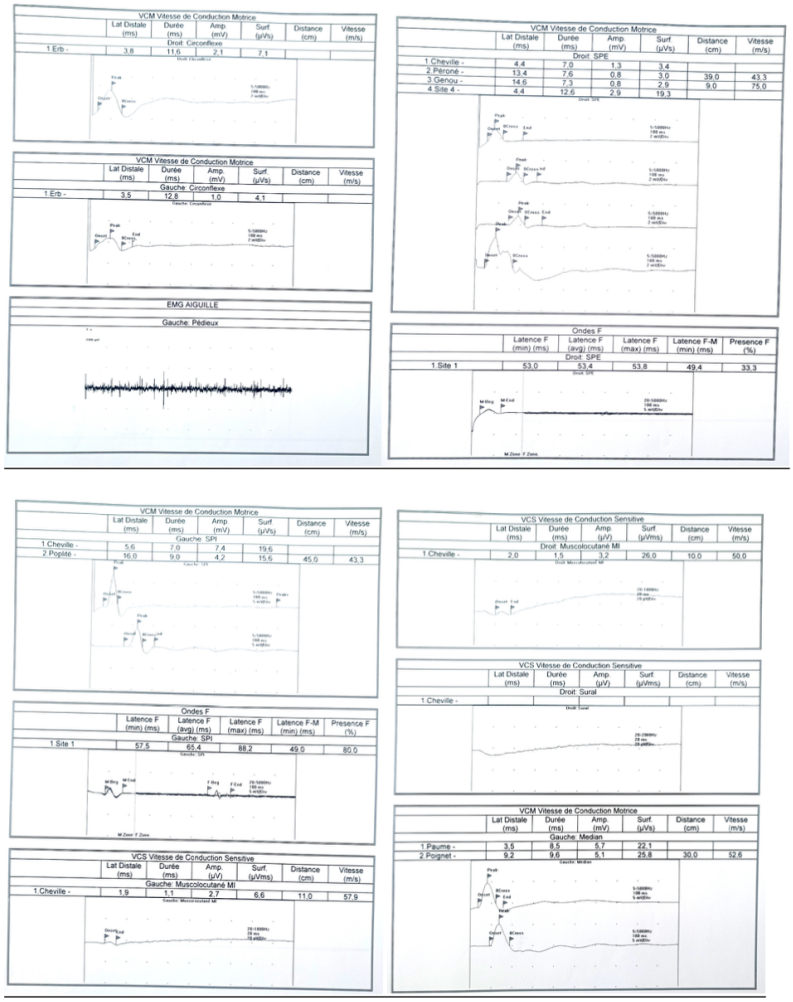

Figure 4. Neurography showing decreased sensory potentials in musculocutaneous nerves and decreased motor potentials in extensor digitorum brevis and anterior tibialis

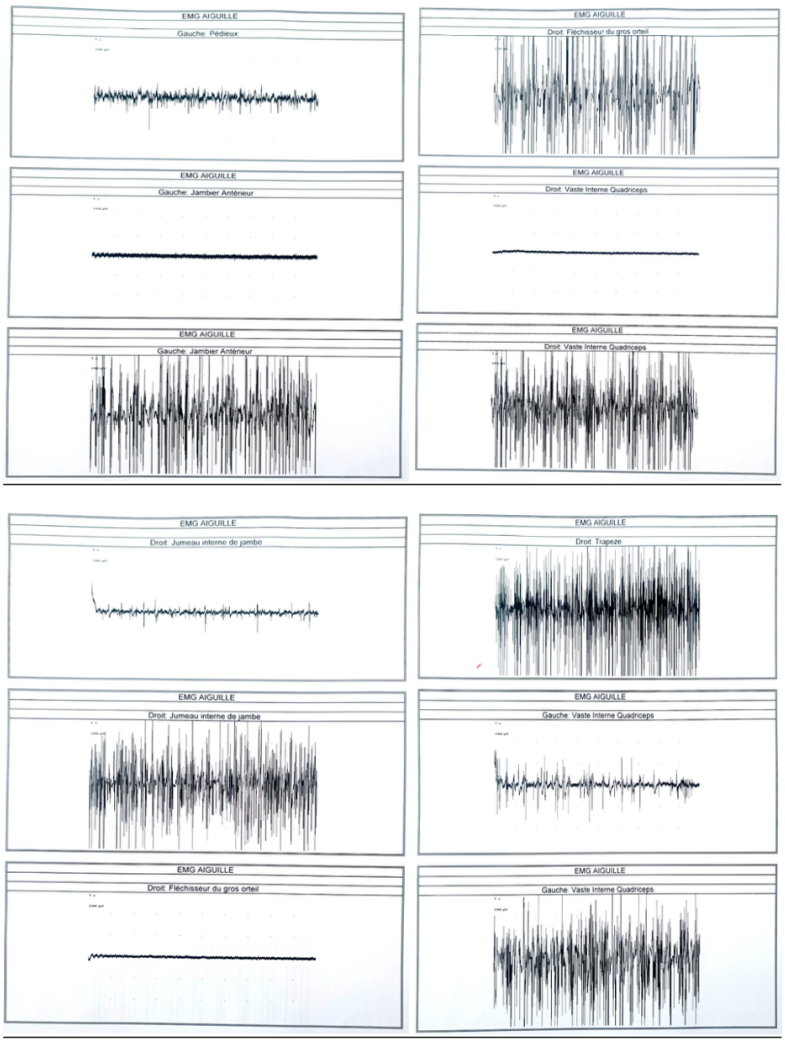

Figure 5. Electromyography showing fibrillation potentials in the left extensor digitorum brevis, right medial gastrocnemius, and left quadriceps

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RESULTATS

EXAMEN	RESULTAT	NORME
HB	8,84	10,0 – 13,0 g/dL
CRP	5,8	0 – 10 mg/L
ASAT (TGO)	66	< 40 UI/L
ALAT (TGP)	63	< 40 UI/L

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Figure 6. Follow-up CPK and LDH results

3. Discussion

Scleromyositis with positive anti-PM75 antibodies is an overlap syndrome between autoimmune myositis and systemic sclerosis. It is a condition that most commonly affects young adults [1, 8].

The clinical manifestations are varied but relatively stereotyped, most frequently involving the muscles, peripheral nervous system, and skin [2, 4]. The clinical features observed in our patient met the 2017 ACR/EULAR diagnostic criteria [10].

In our patient, proximal motor weakness was more severe in the upper limbs. These findings were similar to those reported by Gaudin et al. in 2006, who reported that 10 of their 18 patients had severe proximal muscle weakness [5]. Furthermore, Lorenzo et al. reported that this proximal weakness was more severe in the arm abductors and hip flexors [2].

In addition to peripheral nervous system involvement, extra-neurological manifestations may occur. This was the case in our patient, who presented with both cutaneous and pulmonary involvement [2, 7, 8, 12].

Our patient developed respiratory failure secondary to diaphragmatic involvement. The prevalence of interstitial lung disease associated with systemic sclerosis is 56% (95% CI: 49–63%). Pulmonary involvement is one of the most serious complications and may be life-threatening [11]. Thus, in the EUSTAR cohort, more than 50% of deaths were related to pulmonary involvement [10].

Chest CT may detect interstitial lung disease in up to 90% of patients. These findings were almost identical to those observed in our patient, who presented with respiratory distress and interstitial lung disease on chest CT angiography [5]. Pulmonary CT angiography allowed us to rule out pulmonary embolism, which could have worsened the clinical condition of these patients.

Patients with scleromyositis have serum CPK levels greater than five times the upper limit of normal. This elevation in serum CPK is part of the 2017 ACR/EULAR criteria [10]. These findings were similar to those observed in our patient, whose CPK level was more than nine times the upper limit of normal.

Electromyography is abnormal in approximately 60% of patients with scleromyositis. Spontaneous activity, including positive sharp waves and/or fibrillation potentials, may be observed. Interestingly, a neurogenic pattern may also be found in 25% of patients with scleromyositis [6, 9].

The electroneuromyographic findings in our patient were consistent with those described in the literature, showing myogenic patterns predominantly involving the proximal myotomes, suggestive of polymyositis, associated with a neurogenic pattern [4, 5].

A 2017 meta-analysis reported that 31% of patients with scleromyositis were positive for anti-PM/Scl antibodies [6]. In our patient, anti-PM/Scl antibodies were positive on the myositis DOT assay. However, muscle biopsy could have provided further information regarding muscle ultrastructure [3].

The management of this condition is complex. We initially started treatment with prednisone at 1 mg/kg/day combined with adjunctive therapy, which helped reduce the inflammatory phase and slow respiratory deterioration. Based on the immunological findings, methotrexate 10 mg/week was added, together with folic acid. Methotrexate has also been used in several studies, including that of Marie et al. [11].

The patient also underwent motor rehabilitation sessions. The clinical outcome was favorable, with removal of the nasogastric tube, motor recovery, and a decrease in CPK and LDH levels. However, our patient did not receive intravenous immunoglobulins.

The modified Rankin Scale score at 6 months was 0. This favorable outcome has been described in the literature among patients positive for anti-PM/Scl autoantibodies [5, 7, 11]. Muscle involvement appears to be responsive to immunosuppressive therapy, and patients tend to recover well during the first year of treatment.

4. Conclusion

Scleromyositis is a low-prevalence autoimmune disease that is frequently associated with anti-PM75 antibodies. Some clinical forms may be severe, particularly when the respiratory system is involved. Therefore, rapid diagnosis and multidisciplinary therapeutic management are essential. Corticosteroids and immunosuppressive agents constitute the mainstay of treatment for scleromyositis.

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