




# Nemaline Myopathy in Systemic Lupus Erythematosus

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## Abstract

Sporadic late-onset nemaline myopathy (SLONM) is a rare muscle disorder that can be treated or may pose a potential threat to life. It usually manifests in the later stages of life and is characterized by the accumulation of nemaline rods within muscle fibers. The pathophysiology of this disease is still not fully understood, with some evidence suggesting that autoimmune responses and hematological neoplasia may be involved. Herein, a case of SLONM associated with systemic lupus erythematosus is presented, which showed a favorable response to immunotherapy.

**Keywords:** Anti-signal recognition particle antibody, muscle fibers, nemaline rods, proximal weakness, sporadic late-onset nemaline myopathy, systemic lupus erythematosus

## Presentation

A woman in her 70s presented to the rheumatology clinic with a 2-month history of muscle weakness. She had a long-standing history of systemic lupus erythematosus with lupus nephritis for more than 30 years. Prior to admission, she was on immunomodulatory therapy with oral prednisone 30 mg daily and hydroxychloroquine 200 mg twice daily for elevated urinary protein levels (quantitative urinary protein: 1.39 g/24 h). Physical examination revealed slight to moderate proximal muscle weakness (grade 4/5 in deltoids and grade 3/5 in quadriceps). Serologic studies revealed markedly elevated anti-Smith antibody (>400 RU/mL) and positive anti-dsDNA antibody (217 IU/mL), with concomitant proteinuria of 2.07 g/24 h on quantitative urinalysis. Her anti-signal recognition particle (SRP) antibodies were positive, while creatine kinase levels were normal. Serum protein electrophoresis was normal, and human immunodeficiency virus (HIV) tests returned negative. Electromyography indicated axonal damage in multiple motor fibers of the bilateral lower extremities. Magnetic resonance imaging of the thigh revealed mild muscle edema (Figure 1A, arrows). These clinical and laboratory findings did not align with the typical presentation of SRP-mediated necrotizing myopathy. A quadriceps muscle biopsy was conducted, revealing muscle fibers of varying sizes

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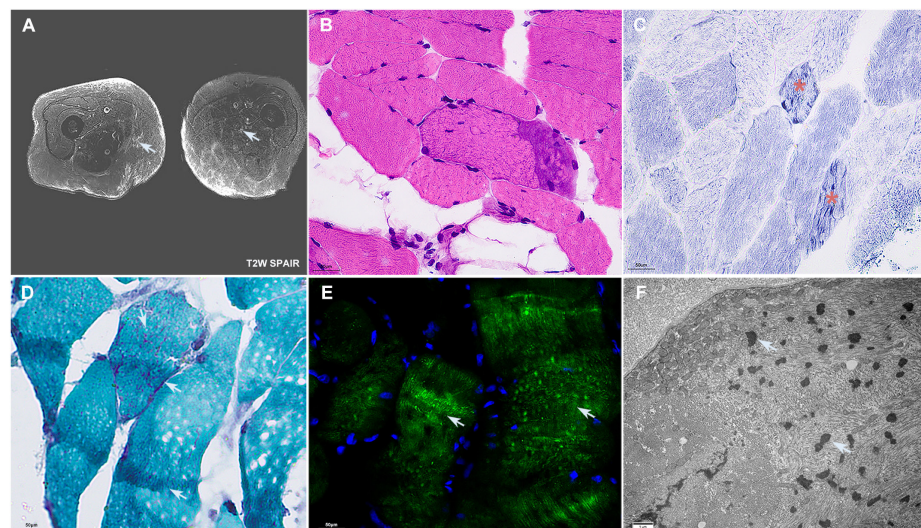
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**Figure 1.** Nemaline myopathy. (A, arrows) Cross-sectional magnetic resonance imaging (spectral attenuated inversion recovery sequence) of the thigh revealed mild muscle edema; (B) Variability in fiber sizes (hematoxylin-eosin stain); (C, asterisks) Irregular internal architecture of muscle fibers (NADH-TR stain); (D, arrows) Dark red-blue structures were visible on modified Gomori trichrome stain; (E, arrows) Actin clustered in regions of muscle fiber cytoplasm (Phalloidin stain); (F, arrows) Electron microscopy showed rod-shaped thickened Z-lines.

(Figure 1B) and the presence of nemaline rods aggregated within the muscle fibers (Figure 1C, asterisks; Figure 1D-F, arrows). Based on these histopathological findings, a diagnosis of sporadic late-onset nemaline myopathy (SLONM) was established. Following a 3-day course of intravenous methylprednisolone (40 mg/day), 7-day immunoglobulin therapy (15 g/day), and a single dose of cyclophosphamide (0.4 g), the patient demonstrated a significant improvement in proximal extremity strength to grade 4+/5 by day 14, with maintained therapeutic response at 3-month follow-up. The patient has given written informed consent to the publication of her case details.

## Discussion

Sporadic late-onset nemaline myopathy is an acquired adult-onset myopathy. It is typically characterized by progressive proximal

muscle weakness and the accumulation of nemaline rods within muscle fibers.<sup>1</sup> Sporadic late-onset nemaline myopathy has been associated with conditions like monoclonal gammopathy and HIV infection.<sup>2</sup> Patients with SLONM usually have normal or only mildly elevated creatine kinase levels.<sup>3</sup> The diagnosis of SLONM mainly depends on identifying nemaline rods in muscle fibers.<sup>1</sup> Under light microscopy, modified Gomori trichrome staining shows these rods as dark red, while Phalloidin staining makes them appear vividly green.<sup>4</sup> There was no indication of inflammation or necrosis observed. Furthermore, vacuolar alterations in the muscle biopsy, which are rarely associated with hydroxychloroquine administration, were absent. Electron microscopy reveals the rods as electron-dense and mostly elongated structures.<sup>4</sup> At present, there is no standardized treatment for SLONM. Various therapeutic approaches have been attempted. These include corticosteroids, steroid-sparing immunosuppressants, immunoglobulins, and chemotherapy.<sup>5</sup> Patients with monoclonal gammopathy-associated SLONM typically exhibit poor prognoses. In contrast, those with autoimmune disease-associated SLONM show significant therapeutic responses to immunomodulatory treatments, correlating with substantially improved clinical outcomes.

**Data Availability Statement:** The data that support the findings of this study are available on request from the corresponding author.

**Ethics Committee Approval:** Ethical committee approval was received from the Ethics Committee of Shanghai Sixth People's Hospital Affiliated to

Shanghai Jiao Tong University School of Medicine (Approval No.: 2025-KY-147(K); Date: 25-Apr-2025).

**Informed Consent:** Written informed consent was obtained from the patients/patient who agreed to take part in the study.

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### Main Points

- Sporadic late-onset nemaline myopathy (SLONM) is a rare disorder typically characterized by the subacute development of weakness.
- Sporadic late-onset nemaline myopathy is pathologically defined by the accumulation of nemaline rods within muscle fibers.
- Sporadic late-onset nemaline myopathy can occasionally be associated with autoimmune disease.
- Immunotherapy appears to improve symptoms of Sporadic late-onset nemaline myopathy.