66-year-old man presented with progressive weakness and myalgia for 3 months, followed by anasarca and dysphagia. Physical examination showed symmetric limb-girdle weakness and generalized edema without skin rashes. Creatine kinase was 1584 U/L (normal: <308). Muscle biopsy displayed inflammatory myopathy with perifascicular pathology (Figure 1). Myositis-specific antibody panel identified positive antinuclear matrix protein 2 (NXP2) autoantibody. Results of a whole-body computed tomography scan were normal except for subcutaneous and soft-tissue edema (Figure 2).

Despite the lack of classic skin rash, the severe diffuse subcutaneous and soft-tissue swelling should raise the suspicion for edematous myositis, which is mostly dermatomyositis. Dysphagia could be from pharyngeal muscle weakness, esophageal calcinosis, or prevertebral soft-tissue swelling, as demonstrated in this patient. Prompt identification of anti-NXP2 antibody in patients with dermatomyositis and these distinct clinical features can serve as useful prognostic markers and guidance for further management.

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FIGURE 1. Hematoxylin and eosin-stained deltoid muscle biopsy. Perifascicular pathology including smallness in size, regeneration, necrosis, internalized nuclei, and vacuolization of fibers at the superficial layer of many fascicles (arrow heads) and perivascular inflammatory cell collections (arrow). Scale bar are 100 μm (A) and 50 μm (B).
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