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CASE REPORT

Spontaneous Retroperitoneal Hemorrhage in Anti-NXP2-Positive Dermatomyositis: a Rare but Severe Complication

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ABSTRACT

Background: **Spontaneous hemorrhage is a rare but serious complication of dermatomyositis (DM).**
Methods: **We report a 61-year-old man with anti-NXP2-positive DM who developed a retroperitoneal hematoma without anticoagulation, shortly after corticosteroid initiation.**
Results: **Laboratory investigations demonstrated rhabdomyolysis, hepatic injury and coagulopathy. The patient was managed with red blood cell transfusion, factor VIII, high-dose corticosteroids and intravenous immunoglobulin, resulting in complete recovery.**
Conclusions: **Although hemorrhagic events in DM are most often linked to anti-MDA5 or anti-Ro52 antibodies, their association with anti-NXP2 antibodies remains uncertain. This case highlights the importance of early recognition and prompt intervention in managing spontaneous hemorrhage in DM.**
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KEYWORDS

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INTRODUCTION

Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by proximal muscle weakness, distinct cutaneous manifestations, and multisystem involvement. Interstitial lung disease (ILD) occurs in 30 - 40% of DM/polymyositis (PM) cases [1], often associated with anti-synthetase antibodies. Rapidly progressive ILD (RP-ILD) is observed in 40 - 79% of patients with anti-MDA5-positive DM and is the primary cause of mortality [2]. Respiratory failure may also arise from weakness in the diaphragm and chest wall muscles. Beyond pulmonary involvement, dysphagia is the most prevalent gastrointestinal symptom, leading to aspiration pneumonia, particularly in elderly patients [3]. Cardiac manifestations, although often subclinical, include conduction abnormalities and a heightened risk of myocardial infarction, which is 2 - 3 times higher in DM/

PM patients [4]. DM is also strongly associated with malignancy, particularly in TIF1γ-positive cases.

Spontaneous intramuscular hemorrhage (SIH) is a rare yet life-threatening complication of DM, characterized by non-traumatic muscle hematomas that can precipitate hemorrhagic shock or coagulopathy, with a mortality rate of 50%. While the pathophysiology of SIH is not fully understood, recent studies indicate that risk factors include male gender, ILD, positivity for anti-MDA5 and anti-Ro52 antibodies, heparin use, intravenous immunoglobulin (IVIG) therapy, and high-dose steroid treatment. This report describes an acute-onset DM case complicated by SIH to highlight the critical need for early recognition and management.

CASE PRESENTATION

A 61-year-old male with a history of chronic alcohol consumption was admitted to our department on January 30, 2025, presenting with a 10-day history of progressive myalgia and symmetric proximal muscle weakness affecting all four limbs. These symptoms developed following a recent upper respiratory infection and were accompanied by a facial and truncal rash, nausea, epigastric discomfort, fatigue, and oliguria. The patient reported no cardiopulmonary symptoms, joint involvement, or photosensitivity. Prior self-administration of ibuprofen and traditional Chinese medicine proved ineffective. Initial laboratory tests revealed a significant increase in creatine kinase levels (11,515 U/L; reference range 53 - 310 U/L).

Physical examination showed tachycardia (118 bpm), hypertension (144/105 mmHg), and a scattered macular rash on the face and upper torso (Supplemental Figure 1). Further laboratory investigations confirmed rhabdomyolysis (lactate dehydrogenase 1,199 U/L; reference range 120 - 250 U/L), hepatocellular injury (aspartate aminotransferase 633 U/L, alanine aminotransferase 189 U/L; reference ≤ 40 U/L), and coagulopathy (activated partial thromboplastin time 42.0 seconds [reference 24.0 - 39.0 seconds], D-dimer 1.73 μg/mL [reference < 1 μg/mL]). Immunological profiling detected anti-NXP2 antibodies (1:100 titer) with weakly positive antinuclear antibodies (1:100 granular pattern), and lymphocyte subset analysis indicated $CD3^+CD4^+$ (296 cells/μL) and $CD3^+CD8^+$ (126 cells/μL) lymphopenia. Additional findings included hyperferritinemia (1,611 ng/mL; reference 4 - 665 ng/mL) and positive fecal occult blood. Serological tests for viral hepatitis, ANCA, anti-cyclic citrullinated peptide antibodies, and respiratory pathogens were negative. Magnetic resonance imaging of the left thigh revealed STIR hyperintensity in the quadriceps, obturator, and pectineus muscles, indicative of acute inflammatory myopathy (Figure 1**)**. Electromyography demonstrated prolonged insertional activity and polyphasic motor unit potentials without evidence of denervation. Chest and abdominal computed tomography (CT) scans showed no signs of ILD.

A diagnosis of anti-NXP2-positive dermatomyositis was confirmed, and treatment with intravenous methylprednisolone (40 mg twice daily) was initiated. Within 72 hours, there was a noticeable improvement in the rash and muscle strength.

However, on day 4, the patient developed acute pain in the left iliopsoas region, which intensified with positional changes. Repeat abdominal imaging revealed an evolving retroperitoneal hematoma (Figure 2) alongside deteriorating coagulopathy (fibrinogen 1.85 g/L, reference 2.0 - 4.0 g/L), D-dimer 6.16 μg/mL, and factor VIII activity 359.2% (reference 60 - 150%). A diagnosis of hemorrhagic myositis with spontaneous retroperitoneal bleeding was made. The management strategy included transfusions of packed red blood cells, supplementation with factor VIII, high-dose corticosteroids (methylprednisolone 1 g/day for 3 days), and intravenous immunoglobulin (0.4 g/kg/day for 5 days). Serial imaging confirmed the resolution of the hematoma by day 10 (Supplemental Figure 2), and the patient's sustained recovery at the 1-month follow-up.

DISCUSSION

Spontaneous hemorrhage is a rare but potentially fatal complication of dermatomyositis (DM), with incidence rates reported between 1 - 2% [5]. Most cases manifest within the acute phase of the disease, typically within six months. Our case report details a unique occurrence of spontaneous hemorrhage in a DM patient with anti-NXP2 antibody positivity, a subtype seldom associated with this complication. This contribution enhances the limited epidemiological data available, broadening the understanding of hemorrhagic events in DM and emphasizing the necessity for increased clinical vigilance, particularly in patients with extensive muscle involvement.

The etiology of spontaneous hemorrhage in DM is thought to be active inflammation that affects both muscles and microvasculature. Hanawa et al. suggest that microvascular inflammation may intensify vascular fragility, predisposing DM patients to intramuscular hemorrhage [6]. This theory corresponds with our patient's clinical presentation, indicating that immune-mediated endothelial damage could play a significant role. Additionally, autoantibodies in different DM subtypes may influence bleeding risk. While anti-MDA5 and anti-Ro52 antibodies are known to be associated with spontaneous hemorrhage, information regarding anti-NXP2-positive DM [5,7] is limited. NXP2 is implicated in transcriptional regulation and RNA metabolism, with its autoantibody positivity associated with severe muscle weakness, calcinosis, and malignancy [8,9]. Although hemorrhage is infrequently reported in anti-NXP2-positive DM, emerging evidence suggests that the coexistence of anti-Ro52 and anti-NXP2 antibodies may increase the risk of severe vascular complications [10]. Further research is required to clarify the role of

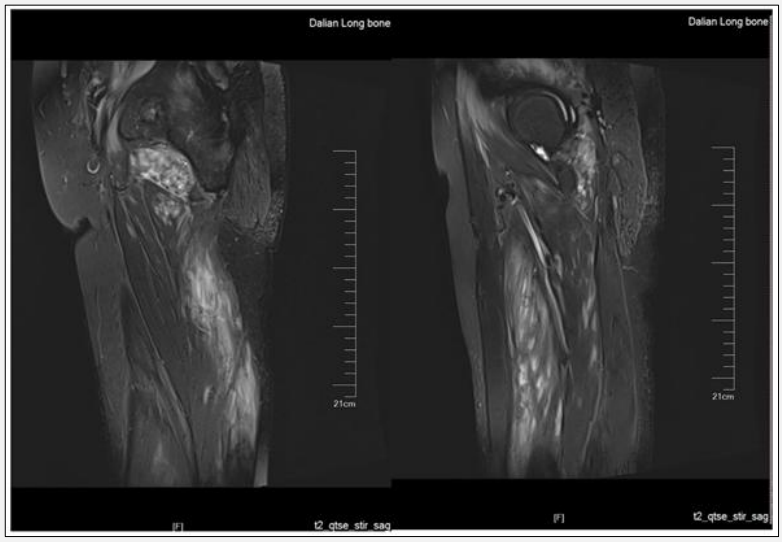

Figure 1. Quadriceps, obturator, and pectineus muscles showed high signal in STIR of left thigh MRI, with edema within affected muscles, subcutaneous and interfascial edema.

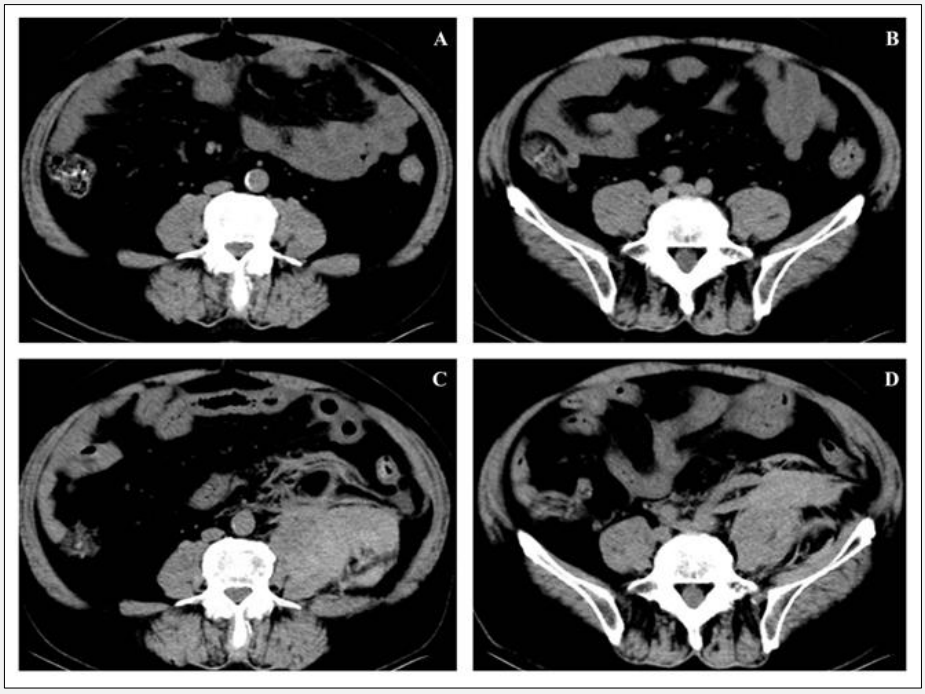

Figure 2. Abdominal CT findings during hospitalization.

Panels A and B show the abdominal CT scan on admission. Panels C and D (day 4) reveal a newly developed left-sided retroperitoneal hematoma adjacent to the iliopsoas muscle.

NXP2 in vascular integrity and bleeding risk.

Early recognition of spontaneous hemorrhage in DM is vital due to its varied clinical presentations, which range from mild, nonspecific muscle pain to life-threatening hemorrhagic shock [10]. Deep muscle hematomas, particularly in the iliopsoas and retroperitoneal regions, are associated with a high mortality risk (60%) compared to superficial hematomas (25%) [10]. In our case, CT identified the hemorrhage, aligning with the literature indicating that CT is the preferred imaging modality for diagnosis (87.5%) [5]. While MRI and ultrasonography (USG) also aid in diagnosis, angiography is particularly valuable in persistent or multifocal bleeding, as it facilitates both localization and embolization therapy [5,11]. The management of antithrombotic therapy in DM patients is controversial, particularly due to its association with increased mortality in those with spontaneous intramuscular hemorrhage (SIH) [6]. More than 50% of the reported hemorrhagic cases occur in patients receiving anticoagulation or antiplatelet therapy, often for deep vein thrombosis prophylaxis [5-7,10-12]. Although thromboprophylaxis is critical due to the immobility-related risks in DM, our case highlights the importance of individualized risk-benefit assessments, particularly for patients with deep muscle involvement. Future research should aim to refine prophylactic strategies that balance the thrombotic and hemorrhagic risks in DM populations.

Spontaneous hemorrhage, while rare, constitutes a severe complication of DM that demands heightened clinical vigilance. Deep muscle hematomas, particularly in the iliopsoas region, pose a substantial mortality risk and require prompt recognition and appropriate diagnostic imaging. Our case highlights the potential involvement of anti-NXP2 antibodies in such hemorrhagic events, highlighting the necessity for further investigation into the immunopathogenesis of vascular complications associated with DM. Future research should explore the role of DM autoantibodies in maintaining endothelial integrity and develop strategies to optimize thromboprophylaxis in patients at high risk.

Consent for Publication:

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images.

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