Acute Kidney Injury Following Dermatomyositis

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Renal involvement in patients with inflammatory myopathies, like dermatomyositis, is rare. We present the case of a woman, aged 66 years, who arrived at the hospital with acute renal failure. She had a diffuse erythematous rash, severe muscle weakness and decreased motor capacity. Inflammatory features of the skin/muscle biopsy along with the dramatic improvement in symptomatology upon the intake of steroids pointed towards dermatomyositis. Recognition of this pattern of events, adopting a multidisciplinary approach, early diagnosis and steroid treatment are necessary for better patient outcomes.

Keywords: Dermatomyositis; Inflammatory myopathy; Kidney injury; Kidney failure; Rhabdomyolysis

Dermatomyositis is a rare autoimmune disease of low prevalence (7 cases/100,000).¹ It is an idiopathic inflammatory myopathy characterized by cutaneous findings and moderate-to-severe muscle weakness.² It is often associated with neoplasia and results from a complement-mediated microangiopathy in the muscle.³ The disease can be associated with the occurrence of various nonspecific myositis-associated antibodies.

Dermatomyositis can lead to frequent and, in some cases, life-threatening extra-muscular complications. In 25% to 45% of cases, pulmonary symptoms can be noted.⁴ Cardiac involvement has also been described in 5% to 15% of cases.⁵ Even though renal involvement is frequent in various systemic autoimmune diseases, the incidence and outcome of renal involvement in patients with inflammatory myopathies remain significantly low.⁶ Dermatomyositis is a rare cause of rhabdomyolysis that occurs because of ongoing destruction of muscle fibers. Being aware of this fact has imperative therapeutic consequences, as patients can develop renal failure. We hereby report a rare case of dermatomyositis presenting with rhabdomyolysis and acute kidney injury.

Case Report

A woman, aged 66 years, presented to the hospital with diffuse erythematous rash, severe muscle weakness, and decreased motor capacity. Prior to her presentation, she had seen other physicians for similar but less severe complaints. She had lower limb edema and diffuse myalgias. Her workup had shown an increased creatinine level reaching 4.6 mg/dL with 24% clearance, and elevated creatine phosphokinase (CPK) levels (1500 U/L). She had increased immunoglobulin G (IgG) levels (2138 mg/dL); however, her IgA/IgG ratio was normal. She also had an elevated level of free light kappa and lambda chains on serum and urine electrophoresis; however, the free kappa/free lambda ratio was normal. A computed tomography (CT) scan of the brain without contrast revealed several hypo-dense lesions in the right and left parietal bone. Furthermore, an autoimmune workup showed a positive anti-Sjogren's syndrome antibody (SSA); however, a biopsy of the salivary gland was negative for Sjogren's syndrome. A renal ultrasound was normal with no signs of arterial stenosis. As the patient was taking a cyclooxygenase-II enzyme (COX-2) inhibitor (200 mg, 1 tablet daily) for low back pain, the previous physicians diagnosed the patient with non-steroidal

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Table 1: Laboratory test values at presentation

Laboratory Test	Patient Value	Normal Levels
Hemoglobin (g/dl)	10.6	12 – 16
MCV (fl)	85	80 – 94
Creatinine (mg/dl)	6.67	0.51 – 0.95
BUN (mg/dl)	123	6 – 20
Potassium (mmol/L)	4.73	3.5 – 5.1
CO ₂ (U/L)	15	23 – 29
Uric Acid (mg/dl)	7.6	2 – 7
Phosphorus (mg/dl)	8.44	2.7 – 4.5
Albumin (g/L)	31	35 – 50
Globulin (g/L)	36	29 – 33
CPK (U/L)	3956	<200
ESR (mm/h)	50	0 – 20
LDH (U/L)	794	135 – 235

MCV, mean corpuscular volume; BUN, blood urea nitrogen; CO₂, carbon dioxide; CPK, creatine phosphokinase; ESR, erythrocyte sedimentation rate; LDH, lactic acid dehydrogenase

anti-inflammatory drug (NSAID)-induced kidney injury with inflammatory repercussions.

Amid symptom progression, the patient was transferred to our medical center. On examination, she had a puffy face with periorbital edema and diffuse erythematous rash. The patient was bedridden, incapable of moving her lower limbs and had absent reflexes. Workup revealed a normocytic anemia and a progressive increase in creatinine and CPK levels (Table 1). The rheumatology team recommended a skin biopsy, which showed evidence of lymphocytic infiltration around vessels at the border of muscle fibers (Figure 1). An electromyogram showed inflammatory myopathy with normal neural conduction, fibrillation at rest, and myogenous trace at effort. A diagnosis of dermatomyositis was suspected.

The patient was started on high-dose corticosteroids with intravenous methyl-prednisone (1 g daily). After 3 days of treatment, the patient was shifted to oral prednisone and azathioprine (50 mg, twice daily), with aggressive intravenous hydration. As a result, creatinine, blood urea nitrogen, and CPK levels decreased progressively back to acceptable limits. Later, a 24-hour urine collection showed mild proteinuria of 280 mg/24 h (normal range: 28-141 mg/24 h). Meanwhile, investigations done to identify the reason behind her kidney injury were normal (Table 2). Further rheumatologic workup

showed that her antinuclear antibody profile was positive for Mi-2 (+), anti-SSA native (++), and Ro-52Ab (+++), which are consistent with dermatomyositis.

The patient was diagnosed with dermatomyositis that induced rhabdomyolysis, which subsequently led to severe acute kidney injury. Marked improvement was noted after initiating corticosteroid treatment for dermatomyositis. When the patient's symptoms subsided, and her creatinine levels were within normal limits, she was discharged.

Discussion

Our patient's presentation was consistent with dermatomyositis, as she exhibited a characteristic erythematous rash, muscle weakness, and electrophysiological recordings suggestive of an inflammatory myopathy. Focal features of muscle inflammation and necrosis along with perifascicular atrophy edged the diagnosis closer towards dermatomyositis. Another potential diagnosis was acute tubular necrosis; however, the development of the characteristic rash, muscular weakness, biopsy results, and the dramatic improvement of symptoms with steroidal treatment pointed to an underlying inflammatory muscle disease, like dermatomyositis.

It is believed that rhabdomyolysis and muscle destruction can result in myoglobinuria. The release of myoglobin from muscle cells causes an increase in levels of CPK and lactic acid dehydrogenase.⁷ When myoglobin is excreted in the

Table 2: Laboratory tests done to determine etiology of nephrological symptoms

Laboratory Test	Patient Value	Normal Levels
Urine culture	No growth	No growth
C3 (g/L)	0.86	0.9 – 1.8
C4 (g/L)	0.27	0.1 – 0.4
HIV (1+2) (S/CO)	0.13	<0.9
HbsAg (S/CO)	0.15	<1
HCV (S/CO)	0.05	<1
Cryoglobulin	Negative	Negative
Anti-cardiolipin IgG	<3	Negative if <12
Anti-cardiolipin IgM	<3	Negative if <12
IgA (g/L)	2.29	0.7 – 4.0
IgG (g/L)	16.36	7 – 16
IgM (g/L)	0.85	0.4 – 2.3

S/CO, signal-to-cutoff; HbsAg, hepatitis B surface antigen; HCV, hepatitis C virus; IgA, immunoglobulin A; IgG; immunoglobulin G; IgM, immunoglobulin M

Figure 1. Infiltration of lymphocytes around vessels at the border of muscle fibers, which is consistent with dermal histological findings in dermatomyositis.

urine, monomers with heme molecules precipitate. This can lead to tubular obstruction and acute renal failure.

A multidisciplinary approach was crucial in reaching the right diagnosis and administering the appropriate treatment. The patient had been hospitalized previously for the same symptoms, but physicians were unable to properly manage the patient. Therefore, collaborative efforts and reasonable debate among physicians and staff from different specialties and backgrounds are necessary for achieving the best outcomes in a patient-centered approach.^{8,9}

Conclusion

Renal involvement in the setting of dermatomyositis is rare; nevertheless, it can occur. Early diagnosis and effective treatment can markedly improve kidney function. Physicians should remain alerted to nephrologic symptoms resulting from dermatomyositis.

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