

Dermatomyositis with renal infarction: a case report and literature review

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Abstract

Renal infarction is a rare clinical entity that is not easily detected by low-sensitivity ultrasound. We herein report a case of dermatomyositis with renal infarction detected during corticosteroid therapy. The patient was followed up for 18 months. A woman who was clinically diagnosed with dermatomyositis complained of severe pain in the right flank of the low back and abdomen, accompanied by nausea and vomiting during corticosteroid therapy. Based on the findings of routine blood tests, abdominal X-ray radiography, and abdominal ultrasound, the patient was diagnosed with acute gastroenteritis and treated with levofloxacin. However, her symptoms were not relieved. Abdominal contrast-enhanced computed tomography revealed renal infarction. Clinicians should be alert to the occurrence of thrombosis, especially when it manifests as vasculitis in patients with rheumatic disease who complain of severe abdominal pain, because it may suggest the presence of renal infarction.

Keywords

Dermatomyositis, renal infarction, vasculitis, Raynaud's phenomenon

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Introduction

Dermatomyositis is an inflammatory myopathy characterized by moderate to severe muscle weakness and inflammation. The skin manifestations include heliotrope rash, Gottron's papules, and cuticular changes. It is also a systemic disorder involving cardiac disturbances, interstitial lung disease, and gastrointestinal ulcerations. General symptoms include fever, malaise, weight loss, arthralgia, and Raynaud's phenomenon. The immunopathology of dermatomyositis begins when putative antibodies directed against endothelial cells activate

complement C3. The complement deposits sequentially lead to swollen endothelial cells, vacuolization, capillary necrosis, perivascular inflammation, ischemia, and destruction of muscle fibers. Macrophages, B cells, and

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CD4 T cells all contribute to the onset of dermatomyositis by secreting cytokines and chemokines. Pathologically, dermatomyositis is a type of secondary vasculitis affecting capillaries.^{1,2} The main treatment is corticosteroids combined with immunosuppressive drugs.

Renal infarction is a rather rare complication that can be caused by obstruction of the renal arterial flow. The most common symptoms include abdominal pain, flank pain, and nausea and vomiting. Many factors may contribute to acute renal infarction, including emboli secondary to cardiac diseases such as atrial fibrillation, atherosclerosis, valvular heart disease, myocardial infarction, ventricular aneurysm, and dilated cardiomyopathy.³ Other causes include trauma, dissection of the renal artery or aneurysm, hypercoagulable states, fibromuscular dysplasia, and idiopathic renal infarction.⁴ The possibility of vasculitis has not been fully addressed. Leukocytosis ($>10 \times 10^9/L$) and an elevated lactate dehydrogenase level ($>620 IU/L$) are the most remarkable laboratory findings, while the kidney function on admission may be normal in most patients.^{5,6} Contrast-enhanced computed tomography (CT) is strongly recommended to diagnose or rule out renal infarction.⁷ We herein present a case involving a female patient with dermatomyositis who developed renal infarction as a complication during corticosteroid therapy.

Case presentation

A 56-year-old woman presented with a 2-month history of swelling and tenderness of the proximal interphalangeal and metacarpophalangeal joints. Two weeks before admission, she had developed progressive symmetric muscle weakness in the proximal extremities accompanied by symptoms of Raynaud's phenomenon and dyspnea after activity. Physical examination upon

admission revealed Gottron's sign and erythema in the orbital cavity, chest, shoulders, and back. Velcro crackles were audible over both lower lung fields. A manual muscle test showed weakness of the muscles (4/5) of her proximal extremities.

Other laboratory tests were unremarkable except for elevated aspartate transaminase (85 U/L), alanine transaminase (82 U/L), lactate dehydrogenase (376 U/L), creatine kinase (CK) (3716 U/L), and erythrocyte sedimentation rate (34 mm/h). Chest CT demonstrated interstitial pneumonia in both lower lung lobes. Electromyography showed myogenic impairment. After excluding the possibility of neoplasia, the patient was diagnosed with dermatomyositis, for which intravenous methylprednisolone was prescribed at 80 mg/day.⁸ The treatment significantly improved her symptoms, and her serum CK level decreased to 1213 U/L.

Two weeks after admission, the patient suddenly complained of severe pain on the right side of the waist, accompanied by nausea and vomiting. Routine blood examination showed a white blood cell count of $25.4 \times 10^9/L$ and a neutrophilic granulocyte concentration of 77%. The serum levels of amylase and troponin were normal. Abdominal ultrasound findings were unremarkable. Abdominal X-ray radiography showed that part of the gut was dilated and a fluid level was present. An oval high-density shadow was simultaneously detected in the left upper quadrant. Gastroenteritis was diagnosed, for which levofloxacin was prescribed. However, the symptoms were not relieved. Subsequent abdominal CT suggested renal infarction in the right kidney (Figure 1a), for which lumbrokinase, aspirin, and atorvastatin were administered.^{9,10} After about 10 weeks of methylprednisolone and cyclophosphamide treatment, the patient's muscle strength in the proximal extremities improved along with a continuous decline in the CK level.

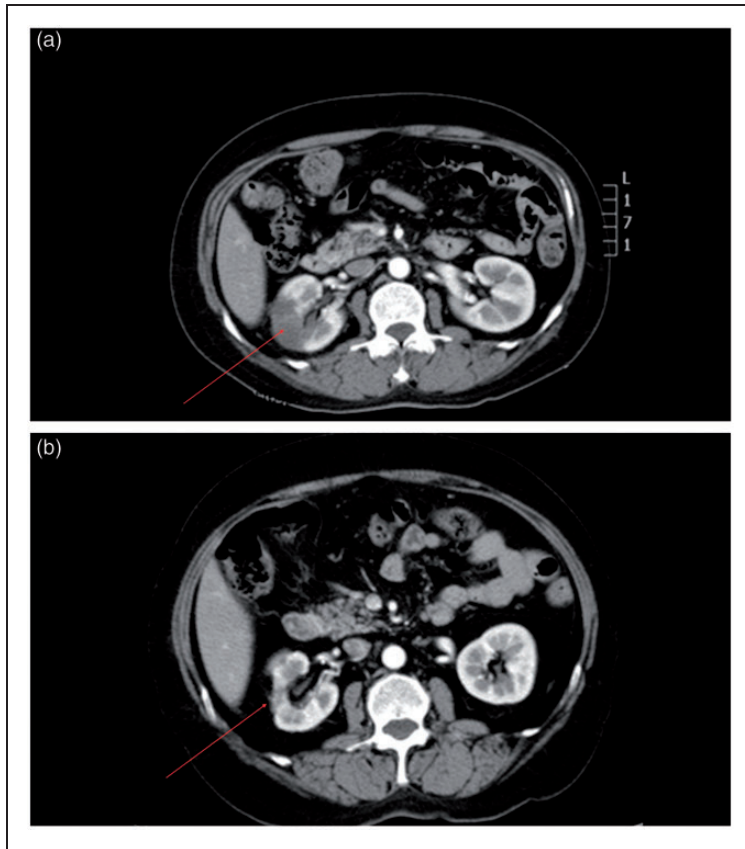


Figure 1. Computed tomography findings. **(a)** Initial contrast-enhanced computed tomography scan of the abdomen shows infarction in the right kidney (arrow). **(b)** Contrast-enhanced computed tomography scan of the abdomen at the 2-year follow-up reveals morphological change of the right kidney (arrow).

By the end of July 2016, the disease course was stable; the prednisolone was tapered to 5 mg/day and the dose of cyclophosphamide was increased to 9.2 g. Above all, although the renal infarction did not seem to have significant impact on serum creatinine level, it caused some morphologic changes in the right kidney (Figure 1b).

Discussion

To the best of our knowledge, only one case of dermatomyositis associated with renal infarction was reported previously.¹¹ The clinical features of that patient and ours are

listed in Table 1. In that case report, a 60-year-old woman was diagnosed with dermatomyositis on the basis of Gottron's sign, a heliotrope rash, elevation of serum myogenic enzymes, and electromyographic and magnetic resonance imaging findings. She was treated with 60 mg/day of prednisolone for 1 week, when she suddenly developed splenic and renal infarctions. Vasculopathy associated with dermatomyositis was the primary contributor to the infarction, which was represented on selective angiography as mild stenosis, interruption, and irregularity of the peripheral arteries of the spleen and left kidney.

Table 1. Clinical features of patients with dermatomyositis and renal infarction

Author	Sex	Age (y)	Symptoms	Infarction sites	Causes	Treatment	Clinical outcome
Matsuda et al. ¹¹	Female	60	Muscle weakness, erythema, heliotrope rash, Gottron's sign, abdominal pain	Left kidney, spleen	Arterial stenosis, interruption, and irregularity	Prednisolone, tissue plasminogen activator, warfarin potassium, CP	CR
Present case	Female	56	Muscle weakness, erythema, heliotrope rash, Gottron's sign, Raynaud's phenomenon, abdominal pain, nausea, vomiting	Right kidney	Not available	MP, lumbrokinase, aspirin, atorvastatin, CP	CR

CP: cyclophosphamide, MP: methylprednisolone, CR: complete remission

The patient was subsequently treated with cyclophosphamide, anticoagulants, and increased doses of corticosteroids and responded well to the treatment.

Patients with dermatomyositis often have a relatively high incidence of arterial events, including acute myocardial infarction, cerebrovascular accidents, and peripheral arterial diseases. Traditional risk factors such as hypertension and dyslipidemia are predictors of arterial events.¹² In the case reported herein, the patient had no other infarction risk factors such as hypertension, hyperlipidemia, or hyperglycemia. The renal infarction in this patient may have been attributed to the following factors. First, the patient had typical Raynaud's phenomenon, which is associated with progressive pathological remodeling within the microvasculature, macrovasculature, and extravascular compartments, resulting in substantial tissue injury.¹³ The increasing vasospastic potential by local cold exposure and stress in the kidney caused renal infarction, resulting in thrombosis and vascular obliteration. This was also reported in a case of renal infarction with polyarteritis nodosa and aneurysmal collapse of the arterial wall.¹⁴ A similar

phenomenon cannot be excluded in our case. Second, the treatment with high-dose corticosteroids might have induced hypertension, hyperlipidemia, and hyperglycemia, which gradually increased the risk of infarction. Meanwhile, the corticosteroids induced a state of hypercoagulability, thus increasing the formation of cell plasminogen activators and inhibiting prostacyclin synthesis.¹⁵ Finally, the inflammatory process may have triggered endothelial dysfunction of small arteries and capillaries. In addition to inflammation, endothelial cells are involved in disturbance of the coagulation system by releasing tissue factors and thrombin inhibitors, ultimately resulting in a procoagulant state.¹⁶

In summary, clinicians should be alert to the occurrence of thrombosis in patients with rheumatic disease, especially when it manifests as vasculitis. When a patient reports severe abdominal pain that cannot be explained by causes commonly encountered in clinical practice, the possibility of visceral thrombosis should be highly suspected. CT is strongly recommended because of the low sensitivity of abdominal ultrasound for renal infarction. Follow-up

involving serial serum creatinine measurements, renal scintigraphy, and measurement of the infarction volume by three-dimensional CT are suggested to assess potential acute and chronic renal function decline.¹⁷

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Declaration of conflicting interests

The authors declare that there is no conflict of interest.

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