Mod Rheumatol (2007) 17:54–56
DOI 10.1007/s10165-006-0529-8

CASE REPORT

**Yoshitaka Morita · Takeo Sakuta · Hajime Nagasu
Atsunori Kuwabara · Yuko Tokuoka · Sanae Teshigawara
Hideyuki Horike · Naoki Kashihara**

Bilateral ureteral stenosis and duodenal perforation in a patient with dermatomyositis

Received: May 29, 2006 / Accepted: September 20, 2006

Abstract We report the case of a 19-year-old man with dermatomyositis who developed abdominal pain and anuria. The examination revealed bilateral ureteral stenosis. The patient also developed multiple ulcerations of the duodenum with perforations. The clinical feature was considered to represent that of juvenile dermatomyositis, which is characterized by systemic necrotizing vasculitis. Rheumatologists should be alerted about this serious complication in patients with childhood or young adult dermatomyositis presenting with abdominal complaints.

Key words Dermatomyositis · Duodenal perforation · Ureteral stenosis

Introduction

Dermatomyositis is defined as an idiopathic inflammatory myopathy with varying cutaneous manifestations. Dermatomyositis presenting in childhood is a disease that is clinically and pathologically distinct from that seen in adults. Juvenile dermatomyositis is typically manifested by necrotizing vasculitis, which involves not only the skin and musculature but often also the gastrointestinal tract.[1] Affected patients may present with abdominal pain, gastrointestinal bleeding, or perforations. Although it is extremely uncommon, the ureter may be involved in childhood dermatomyositis.[2,3] We report an interesting case of young adult dermatomyositis in which bilateral ureteral stenosis and duodenal perforation occurred.

Y. Morita (✉) · T. Sakuta · H. Nagasu · A. Kuwabara · Y. Tokuoka · H. Horike · N. Kashihara
Division of Nephrology and Rheumatology, Department of Internal Medicine, Kawasaki Medical School, 577 Matsushima, Kurashiki 701-0192, Japan
Tel. +81-86-462-1111; Fax +81-86-462-1199
e-mail: morita@med.kawasaki-m.ac.jp

S. Teshigawara
Department of Medicine and Clinical Science, Okayama University Graduate School of Medicine and Dentistry, Okayama, Japan

Case report

A 19-year-old Japanese man in whom polymyositis had been diagnosed 6 months previously was referred to our clinic in April 2004 for recurrence of the disease. He was admitted to the previous hospital in November 2003 because he had fever, systemic muscle pain, and difficulty in standing up. On physical examination, he had pronounced proximal muscle weakness in all four extremities. The chest and abdomen were clinically normal, and skin rash was not observed at that time. Laboratory findings were as follows: C reactive protein (CRP) 0.2 mg/dl, hemoglobin 16.9 g/dl, white cell count 5200 μl^{-1}, and platelets 16.9×10^4 μl^{-1}. Serum creatine kinase (CK) was elevated at 6565 IU/l. Immunological investigations revealed that he had tested negative for Hep-2 antinuclear antibody, anti-Jo-1, anti-double-stranded DNA, anti-Sm, anti-SS-A, and anti-SS-B. Serum complement concentration was normal. Urinalysis was negative for proteinuria and occult blood. The chest X-ray and computed tomography were normal. Elecromyogram revealed the myogenic pattern. Muscle biopsy showed an area of degeneration of myofibers but failed to demonstrate typical findings of inflammatory myopathy or vasculitis. He was clinically diagnosed with polymyositis, which was initially treated with oral prednisolone at a dosage of 55 mg/day. His muscle symptoms improved rapidly with a decrease in the serum CK level to the normal range. However, in April 2004, when tapering the steroid dosage to 30 mg/day, he had experienced an exacerbation of systemic muscle pain with increase of serum CK.

An initial examination at our clinic revealed that he had significant proximal muscle weakness. Skin examination exhibited a diffuse erythematous maculopapular rash over his face and neck. The facial rash involved the upper eyelids and nasolabial area. No rash was observed on the extensor surface of joints. Laboratory tests showed that the white cell count was 12 200 μl^{-1}, CRP 0.06 mg/dl (normal <0.30), CK 900 IU/l (normal <324), and aldolase 18.4 IU/l (normal <6.1). Prothrombin and activated partial thromboplastin times were normal. Antinuclear antibodies, antineutrophil

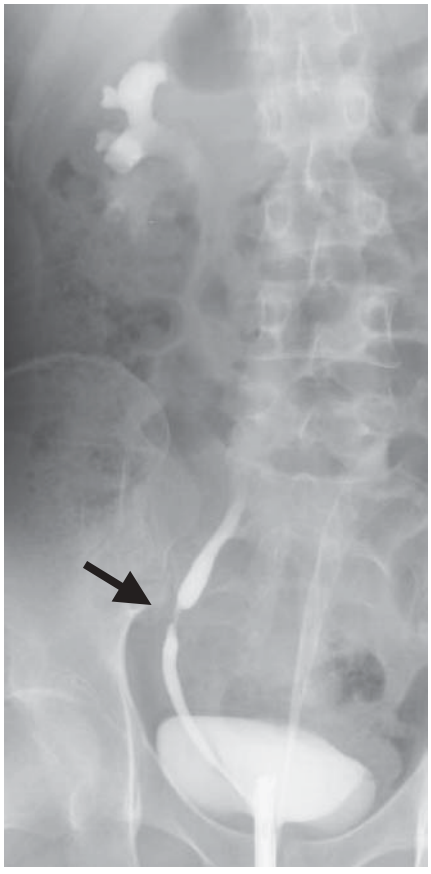

Fig. 1. Retrograde pyelography demonstrating a 1-cm-long stenosis in the right ureter (*arrow*) and hydronephrosis

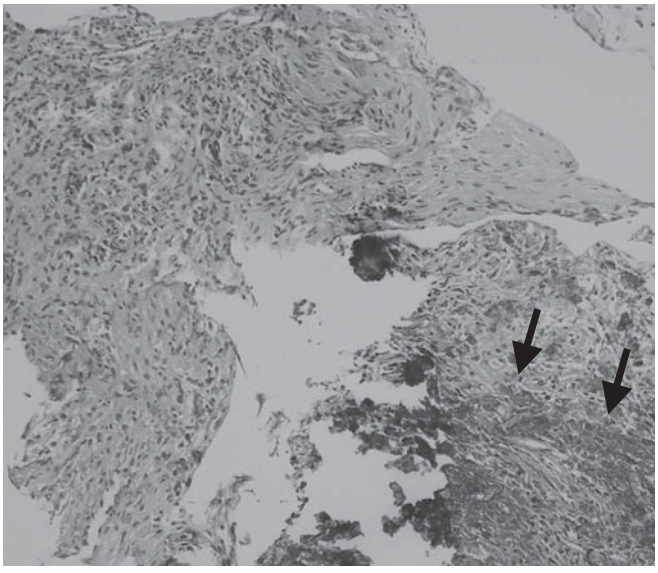

Fig. 2. Biopsy specimen of the stenotic ureter showing calcification (*arrows*) with granulomatous tissue (H&E, ×*200*)

cytoplasmic antibodies, anticardiolipin antibodies, and lupus anticoagulants were all negative. The cutaneous findings associated with myositis led to an alternative diagnosis of dermatomyositis. The dosage of prednisolone was increased to 60 mg/day. The muscle pain rapidly resolved and the serum CK level decreased to the normal range.

Six weeks after his initial presentation, the patient complained of pain in the right flank. The serum CRP level was transiently elevated at 1.08 mg/dl. White cell count was 6840 μl^{-1} and CK 153 IU/l. Urinalysis was negative for occult blood. A computed tomographic scan demonstrated right hydronephrosis, and retrograde pyelography showed a 1-cm-long stenosis in the right lower ureter (Fig. 1). Histopathological examination of a specimen obtained from the stenotic ureter at endoscopic biopsy showed calcification with granulomatous tissue (Fig. 2). No vasculitic lesion was observed in the tissue. A few weeks later, he developed anuria because of the appearance of an obstruction of the left side of the urinary tract. Exploration revealed a 1-cm-long obstruction of the left lower ureter. Stents were successfully inserted bilaterally into each of the stenotic ureters. The steroid dosage was gradually tapered without the recurrence of myositis.

However, in November 2004, he presented with intractable abdominal pain, while taking prednisolone 15 mg/day and famotidine 20 mg/day for prevention of gastroduodenal ulcer. Laboratory findings were CRP 12.2 mg/dl and white cell count 14 900 μl^{-1}. Plain radiographs revealed intestinal perforation. An emergency laparoscopic operation showed an ulcer and perforation at the second portion of the duodenum, and repair of the perforation was undertaken. The postoperative course was uneventful. Gastrointestinal endoscopy after the operation showed multiple ulcerations of the second and the third portions of the duodenum. Although histological evidence was not obtained, the unusual site of duodenal ulcer suggested active intestinal vasculitis. The dosage of prednisolone was increased to 40 mg/day. Omeprazole, a proton pump inhibitor, was also given at 40 mg/day. A subsequent endoscopy performed a few weeks later showed that the multiple duodenal ulcers had significantly improved. He was discharged from the hospital in good health. In December 2005, the ureteral stents were removed. He remains clinically well, taking prednisolone 15 mg/day at the time of this report.

Discussion

There are some clinical differences between juvenile and adult forms of dermatomyositis. For example, gastrointestinal vasculopathy is commonly described in juvenile dermatomyositis, but it is rare in adult dermatomyositis.[1] By definition, onset of juvenile dermatomyositis occurs in children younger than the age of 16 years. Our patient with dermatomyositis, who was 19 years old at the onset, was considered to have systemic vasculopathy involving the urinary and intestinal tracts, which may be present as features of juvenile dermatomyositis. Pulmonary disease, which occurs much less frequently in juvenile dermatomyositis, was not observed in our patient. Other clinical manifestations of juvenile dermatomyositis resemble those of adult derma-

tomyositis, including symmetric proximal muscle weakness, myalgias, muscle tenderness, fever, and rash.

Ureteral stenosis complicating dermatomyositis (or juvenile dermatomyositis) is very rare. According to the available published reports, there are only two cases of dermatomyositis with bilateral ureteral stenosis.[2,3] The previous cases occurred in childhood. A vascular mechanism is possibly associated with this complication because of the bilateral symmetry and the relative lack of blood flow in the involved region. Indeed, in one of the reported cases with juvenile dermatomyositis, vasculitis with ureteral necrosis was demonstrated by histopathological examination at autopsy.[3] Ureteral stenosis has been reported in other forms of small-vessel vasculitis such as Wegener's granulomatosis,[4–6] polyarteritis nodosa,[7,8] and Schönlein–Henoch purpura.[9] It is thus reasonable to consider that abdominal vasculopathy in juvenile dermatomyositis leads to ureteral stenosis as in other forms of vasculitis.

Of note, our case developed duodenal perforation as well as ureteral stenosis. Duodenal perforation in this disease has been reported previously.[10–13] This complication can be life-threatening. Two of the six patients with duodenal perforation died because of severe infection.[10–13] In our patient, multiple ulcers were found from the second to the third portions of the duodenum, with perforation in the second portion. The unusual site of duodenal ulcer and perforation is consistent with findings in the previous reports.[10–13]

The exact etiology of abdominal vasculopathy in this disease remains unclear. Therefore, there is no diagnostic laboratory test for this condition or no certain clinical marker to assess the disease activity. Indeed, serum levels of CRP or muscle enzymes were not directly correlated with the disease activity of ureteral or intestinal involvement, at least in our patient. Several studies have demonstrated that an elevated concentration of plasma von Willebrand factor is associated with vascular damage in children with dermatomyositis.[14,15] Although we did not measure it in our case, this test might be useful for monitoring disease activity in some patients.

Dermatomyositis presenting in childhood is characterized by systemic vasculopathy which may lead to ureteral stenosis and duodenal ulcer with perforation. The unique involvement may hamper the diagnosis in the absence of classic vasculitis locations such as kidney, lung, and nerve. Rheumatologists who treat young adults should be aware of this serious complication of dermatomyositis.

References

1. Banker BQ, Victor M. Dermatomyositis (systemic angiopathy) of childhood. Medicine (Baltimore) 1966;45:261–89.
2. Le Guillou M, Richard F, L'Henaff F, Ferriere JM, Durand J, Lacert P, et al. Bilateral ureteral necrosis in a child with dermatomyositis. Eur Urol 1980;6:190–1.
3. Borrelli M, Prado MJ, Cordeiro P, Wroclawski ER, Monteiro Junior J, Kiss MH, et al. Ureteral necrosis in dermatomyositis. J Urol 1988;139:1275–7.
4. Kamar N, Malavaud B, Alric L, le Mao G, Mazerolles C, Duffaut M, et al. Ureteral stenosis as the sole manifestation of Wegener's granulomatosis. Urology 2003;62:352.
5. Plaisier EM, Mougenot B, Khayat R, Gattegno B, Mignon F, Ronco PM. Ureteral stenosis in Wegener's granulomatosis. Nephrol Dial Transplant 1997;12:1759–61.
6. Huong DL, Papo T, Piette JC, Wechsler B, Bletry O, Richard F, et al. Urogenital manifestations of Wegener granulomatosis. Medicine (Baltimore) 1995;74:152–61.
7. Samellas W, Bellonias E, Papacharalampous N. Polyarteritis nodosa: ureteral involvement. J Urol 1971;105:186–7.
8. Kaskarelis IS, Zarifi M, Dantis P, Vrachliotis G. Bilateral ureteral involvement in polyarteritis nodosa. Scand J Urol Nephrol 1995;29:323–6.
9. Pfister C, Liard-Zmuda A, Dacher J, Dubois D, Grise P, Mitrofanoff P. Total bilateral ureteral replacement for stenosing ureteritis in Henoch–Schönlein purpura. Eur Urol 2000;38:96–9.
10. Kaplinsky N, Hod C, Gal-Semo R, Frankl O. Spontaneous duodenal perforation during fulminant dermatomyositis. J Am Med Womens Assoc 1978;33:213–4.
11. Magill HL, Hixson SD, Whitington G, Igarashi M, Hannissian A. Duodenal perforation in childhood dermatomyositis. Pediatr Radiol. 1984;14:28–30.
12. Schullinger JN, Jacobs JC, Berdon WE. Diagnosis and management of gastrointestinal perforations in childhood dermatomyositis with particular reference to perforations of the duodenum. J Pediatr Surg 1985;20:521–4.
13. Wang IJ, Hsu WM, Shun CT, Chiang BL, Ni YH. Juvenile dermatomyositis complicated with vasculitis and duodenal perforation. J Formos Med Assoc 2001;100:844–6.
14. Guzman J, Petty RE, Malleson PN. Monitoring disease activity in juvenile dermatomyositis: the role of von Willebrand factor and muscle enzymes. J Rheumatol 1994;21:739–43.
15. Bloom BJ, Tucker LB, Miller LC, Schaller JG. Von Willebrand factor in juvenile dermatomyositis. J Rheumatol 1995;22:320–5.