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CASE REPORT

Dermatomyositis with hemorrhagic myositis

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Abstract A 64-year-old Japanese female, diagnosed as dermatomyositis with acute interstitial pneumonia, complained of acute abdominal pain. Computed tomography of the abdomen showed hematoma in the right retroperitoneum and left rectus-sheath. Angiogram showed multiple small aneurysms on left iliolumbar artery and a horizontal linear flush, suggesting active bleeding foci in the muscles. Although arterial embolization therapy was effective for hemostatic treatment, she died of thrombotic thrombocytopenic purpura and multiple organ failure without respiratory insufficiency. Other causes of microaneurysm, such as systemic vasculitides or infectious diseases, were excluded. We considered that this is the first case report of dermatomyositis with hemorrhagic myositis associated with small aneurysms.

Keywords Dermatomyositis · Hemorrhagic myositis · Small aneurysm

Introduction

Dermatomyositis is an inflammatory myopathy with characteristic skin lesions, typically heliotrope (slightly cyanotic erythema) on the malar and supraorbital areas of the face, and other areas including the shoulders, upper back, and upper arms [1]. Dermatomyositis may be associated with systemic manifestations including restrictive and interstitial lung disease and cardiomyopathy. The inflammatory mechanism is attributed to a primary T-helper-cell dependent, B-cell-mediated, local humoral immune mechanism, which causes vascular occlusion and capillary obliteration with ischemic change in the skin and muscle [1, 2]. There have been a few reports of dermatomyositis causing hemorrhagic myositis [3, 4]. We present a patient being treated for dermatomyositis, complicated acute abdominal pain resulting from spontaneous hemorrhage in the right psoas and iliacus muscles and the left rectus-sheath. Angiogram during selective catheterization showed multiple small aneurysms and active bleeding foci in the muscles. To the best of our knowledge, this is the first case report of dermatomyositis with hemorrhagic myositis associated with small aneurysms. Spontaneous abdominal hematoma by hemorrhagic myositis may be a cause of acute abdominal pain in patients with dermatomyositis.

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Case report

The patient is a 64-year-old Japanese woman. She had worked as a school nurse until age of 60. Her medical and her family histories gave no abnormal findings without smoking history. On January 2007, skin erythema with a low-grade fever appeared. She developed progressive skin lesions, high fever, dry cough, and dyspnea, and was admitted to our hospital for examination in May 2007. On admission, her height was 156.2 cm, body weight 48.5 kg, and body temperature 37.2°C. Her blood pressure was 109/68 mmHg, pulse rate 83 per minute with a regular rhythm, and respiration 20 breaths per minute. No jaundice or anemia was seen in conjunctiva bulbi or palpebrae, respectively. Cardiovascular system was unremarkable. Ausculation of her chest revealed basal fine crackling

sounds in both lungs. The abdomen was soft and flat. Neither the liver nor the spleen was palpable. Manual muscle testing for the biceps and quadriceps muscles showed grade 4–5, and muscle pain was not present. Although, heliotrope on the supraorbital areas of the face was not apparent, scaly erythema on the dorsum of the hands (Gottron's sign) and skin erythema on the shoulders, upper back, and upper arms were observed. Raynaud's phenomenon and joint swelling were not observed. In neurological examination, pathological reflex was negative.

Laboratory studies on admission to our hospital were as follows. The urine and a complete blood count were normal. The titer of antinuclear antibodies (ANA) was slightly increased at 23×, and the serum level of creatine phosphokinase and aldolase was also slightly elevated at 432 IU/l (normal, 45 to 163), and 11.5 IU/l (normal, 1.7 to 5.7), respectively. The lactate dehydrogenase was elevated at 531 IU/l (normal, 119 to 229), as were aspartate aminotransferase at 320 IU/l (normal, 13 to 33), and were alanine aminotransferase at 136 IU/l (normal, 6 to 27). In addition, the serum KL-6 level was 1,172 U/ml (cutoff level, 500). Anti-U1-RNP, anti-DNA, anti-Jo-1, and proteinase-3 (PR3) or myeloperoxidase (MPO) anti-neutrophil cytoplasmic antibodies (ANCA) were all negative. Antibodies against syphilis by using rapid plasma reagin (RPR) and Treponema pallidum hemagglutination assay (TPHA) were both negative. The cultures of sputum, urine, or blood were all negative. Arterial blood gas analysis showed a decrease in arterial partial pressure of oxygen (73.9 Torr at O_2 3 l/min through nasal cannula). The chest X-ray film shows reticular shadows with basilar predominance (Fig. 1a). High-resolution computed tomography (HRCT) revealed that ground-glass opacities, irregular reticular opacities, and consolidation involving predominantly the lower lung zones (Fig. 1b–e). These radiographical findings were rapidly progressive for a few weeks. An electromyogram of left biceps muscles showed a very slight evidence of increased membrane instability and myopathic features.

On the basis of these findings, we diagnosed this case as clinically amyopathic (or hypomyopathic) dermatomyositis with a rapidly progressive interstitial pneumonia [5, 6]. Intravenous methylprednisolone (1,000 mg/day for 3 days) was given, followed by a combination therapy with oral prednisone at 50 mg/day, oral cyclosporin A at 150 mg/day, and intravenous pulse cyclophosphamide at 500 mg every 2 weeks. Treatment with prophylactic low-molecular-weight heparin 3,750 IU/day (75 IU/kg/day of dalteparin) was started being calculated from the patient's weight as 50 kg.

After 9 days of combination treatment, the patient's respiratory insufficiency was not deteriorated. However, on day 10 she complained of a lower abdominal pain of rapid onset. The CT of the abdomen showed hemorrhage in the right psoas and iliacus muscles and the left rectus-sheath (Fig. 2). Her hemoglobin had fallen from 10.7 to

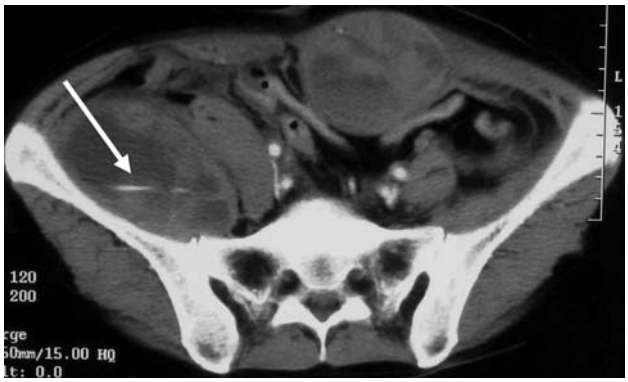

Fig. 2 An enhanced CT scan of the abdomen on day 10. Abdominal CT scan shows massive hemorrhage in the left rectus abdominous and right retroperitoneal space. Note that a horizontal line in the retroperitoneal space suggesting fresh arterial bleeding (*arrow*)

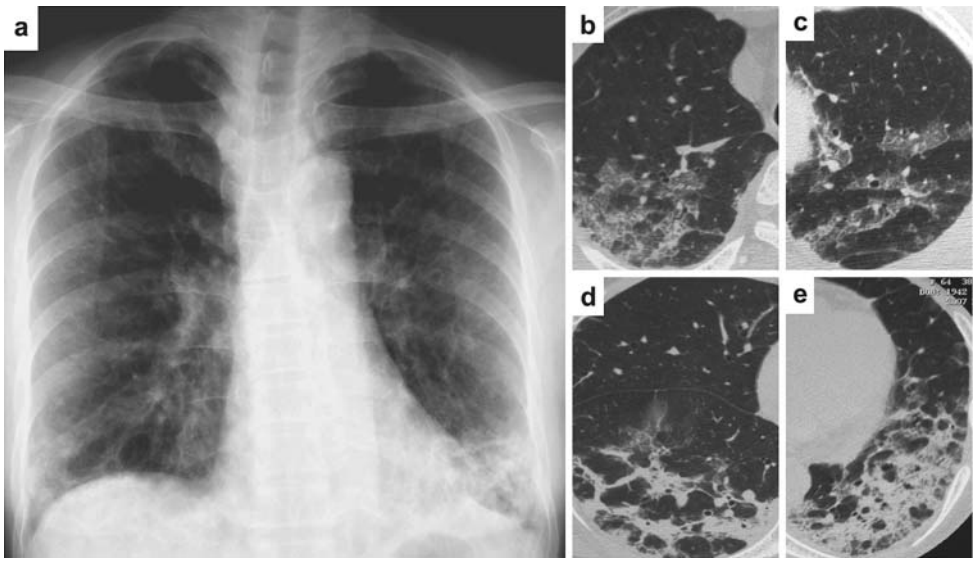

Fig. 1 Lung radiological images on admission. **a** Chest radiograph shows reticular shadows with basilar predominance. **b–e** HRCT (**b**, *right upper* lobe; **c**, *left upper* lobe; **d**, *right middle* and *lower* lobes; and **e**, *left lower* lobe) revealed that ground-glass opacities, irregular reticular opacities, and consolidation involving predominantly the lower lung zones

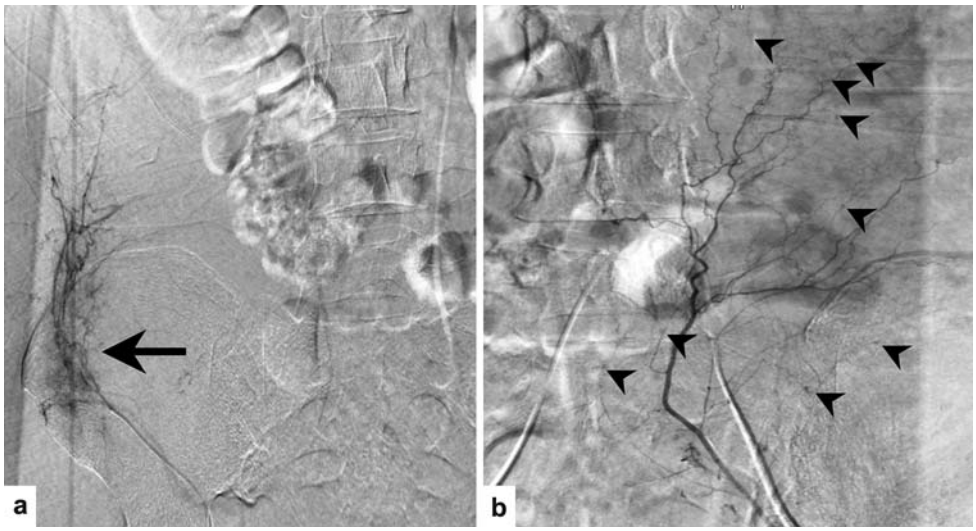

Fig. 3 Angiography findings on day 10. **a** Bleeding from the *right* deep iliac circumflex arteries and iliolumbar artery with iliopsoas hematoma are shown (*arrow*). **b** Bleeding from the *left* inferior epigastric arteries with rectus-sheath hematoma and multiple small aneurysms on left iliolumbar artery are shown (*arrow heads*)

5.5 g/dl overnight, but the platelets were within the reference range. The activated partial thromboplastin time (APTT) and prothrombin time (PT) were not prolonged. The dalteparin was stopped and 3 units of packed red blood cells were transfused. Subsequently transfemoral diagnostic angiography was performed. Digital substraction angiography of the aorta and pelvic arteries was first performed to localize the bleeding site, followed by selective catheterization for detailed diagnostic and therapeutic purposes, immediately. On angiography, active bleeding from the right deep iliac circumflex arteries, iliolumbar artery with iliopsoas hematoma, and from the left inferior epigastric arteries with rectus-sheath hematoma was demonstrated at the corresponding sites of the CT findings (Fig. 3a). Furthermore, left iliolumbar artery had multiple small aneurysms (Fig. 3b). Selective arterial embolization was performed, using gelatine sponge (Spongel; Astellas Pharma Inc., Tokyo, Japan) and an additional microcoil (Hilal embolization micro coil; Cook Inc., Bloomington, IN, USA). Embolization led to rapid hemodynamic stabilization with no complications related to the embolization procedure. Embolization of these vessels prohibited the bleeding both angiographically and clinically for 1 week. The other causes of microaneurysm, such as systemic vasculitis or infectious diseases, were excluded because ANA, MPO-ANCA, PR3-ANCA, RPR, TPHA, and culture of blood were all negative. We considered this dermatomyositis case with hemorrhagic myositis associated with small aneurysm. Although we had stopped anti-coagulation therapy, she recurred bleeding from the same sight on day 17. Embolization were performed again and succeeded, but she died of thrombotic thrombocytopenic purpura and multiple organ failure without respiratory insufficiency on day 28.

Discussion

Hemorrhagic myositis is not a recognized complication of dermatomyositis or side effects of its treatment. In adults, retroperitoneal hemorrhage is most commonly due to rupture of an abdominal aortic aneurysm, but may also be caused by other abdominal conditions, especially renal and adrenal disease (including tumors, renal parenchymal disease, spontaneous rupture of an otherwise normal kidney, and adrenal apoplexy) [7]. Systemic conditions including pregnancy, coagulation disorders, vasculitis (especially polyarteritis nodosa), and renal artery aneurysms or malformations also cause retroperitoneal hemorrhage [7]. The clinical presentation is usually an acute or subacute onset including the features described in our patients. Diagnostic imaging with ultrasound and CT scan of the abdomen is useful to identify the site of the hemorrhage and to define its cause. Treatment should be dependent upon the causes revealed by these investigations [8].

To the best of our knowledge, there are only three case reports of hemorrhagic myositis associated with dermatomyositis [3, 4]. We summarized these four patients including our case (Table 1). Case 1: a 50-year-old woman was diagnosed as having a massive rectus abdominis hematoma associated with dermatomyositis and extensive calcification of the abdominal wall. She was transfused a total of 4 units of packed red blood cells, and over 4 weeks the hematoma resolved spontaneously. Her dermatomyositis remained stable on prednisone 17 mg and azathioprine 100 mg daily, and intravenous immunoglobulin 30 g every 6 weeks. She had no recurrent hemorrhage [3]. Case 2: an 11-year-old girl was diagnosed as a spontaneous retroperitoneal hematoma associated with dermatomyositis. Cause of the hemorrhage was uncertain. She made a good recovery with

Table 1 Mustle hematoma in dermatomyositis

Case	Age	Gender	Bleeding sites	Cause	Heparin	Therapy	Outcome	References
1	50	F	Left rectus abdominis	Calcification?	No	Transfusion	Alive	[3]
2	11	F	Right retroperitoneum	Unknown	No	None	Alive	[3]
3	80	M	Left rectus sheath and oblique, right thigh	Heparin	Yes	Transfusion	Alive	[4]
4	64	F	Right retroperitoneum, left rectus sheath	Aneurysm	Yes	Embolization	Death	Our case

resolution of the pain within a few days. She had no further hemorrhage on prednisone 2 mg/day for 2 years [3]. Case 3: an 80-year-old man diagnosed as dermatomyositis was treated by intravenous methylprednisolone followed by oral prednisone. Treatment with prophylactic unfractionated heparin was also started subcutaneously. Although his APTT, PT, and bleeding time were normal, he suffered from abdominal hemorrhage in the rectus-sheath and oblique muscles. The subcutaneous heparin was stopped and 3 units of packed red blood cells and 4 units of fresh frozen plasma were transfused. The patient successfully recovered and returned home [4].

Although our patient received dalteparin at the first bleeding event, she was not infused dalteparin at the second bleeding. Furthermore, we demonstrated small aneurysms and active bleeding foci in the muscles. Therefore, we considered this case dermatomyositis with hemorrhagic myositis associated with small aneurysm. Because we could not perform autopsy, we were not able to obtain more detailed information. Recently, Sugimoto et al. [9] have shown that an adult patient with clinically amyopathic dermatomyositis with interstitial lung disease complicated thrombotic microangiopathy. Our present case is also amyopathic dermatomyositis with interstitial lung disease. The relationship between small aneurysms and dermatomyositis is unknown, but we speculated that microangiopathy by dermatomyositis might occur in the muscles through T-helper-cells, B-cells, and local humoral immune mechanisms, or the possibility of potential complication of polyarteritis nodosa.

In conclusion, this is the first case report of dermatomyositis with hemorrhagic myositis associated with small aneurysms. Abdominal hematoma by hemorrhagic myositis may be a cause of acute abdominal pain in patients with dermatomyositis. The association of small aneurysms and dermatomyositis should be further examined.

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References

1. Bohan A, Peter JB (1975) Polymyositis and dermatomyositis. N Engl J Med 292:344–347
2. Emslie-Smith AM, Engel AG (1990) Microvascular changes in early and advanced dermatomyositis: a quantitative study. Ann Neurol 27:343–356. doi:10.1002/ana.410270402
3. Orrell RW, Johnston HM, Gibson C, Cass RM, Griggs RC (1998) Spontaneous abdominal hematoma in dermatomyositis. Muscle Nerve 21:1800–1803. doi:10.1002/(SICI)1097-4598(199812)21:12<1800::AID-MUS29>3.0.CO;2-Z
4. Langguth DM, Wong RC, Archibald C, Hogan PG (2004) Haemorrhagic myositis associated with prophylactic heparin use in dermatomyositis. Ann Rheum Dis 63:464–465. doi:10.1136/ard.2003.010272
5. Euwer RL, Sontheimer RD (1993) Amyopathic dermatomyositis: a review. J Invest Dermatol 100:124S–127S. doi:10.1111/1523-1747.ep12356896
6. Suda T, Fujisawa T, Enomoto N, Nakamura Y, Inui N, Naito T, Hashimoto D, Sato J, Toyoshima M, Hashizume H, Chida K (2006) Interstitial lung diseases associated with amyopathic dermatomyositis. Eur Respir J 28:1005–1012. doi:10.1183/09031936.06.00038806
7. Swift DL, Lingeman JE, Baum WC (1980) Spontaneous retroperitoneal hemorrhage: a diagnostic challenge. J Urol 123:577–582
8. Pode D, Caine M (1992) Spontaneous retroperitoneal hemorrhage. J Urol 147:311–318
9. Sugimoto T, Hajiro T, Fujimoto T, Kojyo N, Horie M, Kashiwagi A (2007) Thrombotic microangiopathy in an adult patient with clinically amyopathic dermatomyositis complicated with interstitial lung disease. Lupus 16:1004–1005. doi:10.1177/0961203307082382