Rheumatol Int (2013) 33:2949–2950
DOI 10.1007/s00296-012-2492-3

LETTER TO THE EDITOR

A case of dermatomyositis accompanied by spontaneous intramuscular hemorrhage despite normal coagulability

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Received: 16 February 2012 / Accepted: 13 July 2012 / Published online: 4 August 2012

Dear Editor,

A 60-year-old man presented with a 1-month history of high fever and an erythematous rash on the back and trunk. Gottron's papules and mechanic's hand lesions were also observed. Muscle weakness was not detected by manual muscle testing. Laboratory examination showed elevations of creatine kinase (807 IU/l; 64–279) and aldolase (10.9 U/l; 2.1–6.1). Coagulation test was normal. Antinuclear antibody test result was positive, with a speckled pattern, and titer 1:80. Anti-DNA, anti-Jo-1, and anti-U1-RNP antibodies were negative. High-resolution computed tomography (HRCT) did not detect any internal organ abnormalities. An electromyogram of the muscles was normal. A muscle biopsy showed sparse infiltration of lymphocytes between the muscle fibers. On the basis of these findings, dermatomyositis was diagnosed. Administration of oral prednisolone (60 mg/day) was started. On day 25, patient was in respiratory failure. HRCT detected consolidation and ground glass opacities. Therefore, an i.v. pulse of methylprednisolone (1,000 mg/day for 3 days) was initiated. Unfractionated heparin (UFH) (10,000 IU) was also started to prevent venous thrombosis. After this treatment, the patient had 2 episodes of spontaneous intramuscular hemorrhage. The first episode occurred after 6 days of pulse methylprednisolone treatment. The patient complained of rapid-onset back pain. A CT revealed intramuscular hemorrhage in the left deltoid with hematoma spread into the intra fascia (Fig. 1a). Activated partial thromboplastin time [APTT (89.2 s; normal range, 24.8–35.0)] was prolonged, and UFH administration was stopped. However, 3 days after the first hemorrhage, a second episode occurred despite normal coagulability. The patient complained of sudden cervical pain and a 12-cm subcutaneous hematoma was revealed with no injury detected. A CT demonstrated another intramuscular hematoma in the trapezius muscle (Fig. 1b). The patient's hemoglobin had fallen from 97 to 67 g/l; coagulation parameters were all within normal ranges. The autoantibodies related to coagulation disorder were not detected. The patient needed a transfusion (2 units of packed red blood cells). The patient subsequently experienced an exacerbated IP and died. Dermatomyositis accompanied by spontaneous intramuscular hemorrhage is neither a recognized complication of dermatomyositis nor a side effect of its treatment. There have been 6 other reported cases of dermatomyositis accompanied by spontaneous intramuscular hemorrhages [1–5] (Table 1). In case 5, the patient had also received i.v. UFH, but APTT was normal [5]. The patients in cases 2 and 3 had not received any anticoagulant therapy [2]. In our case (no. 7, Table 1), the patient was treated with i.v. UFH and methylprednisolone. Therefore, there is a possibility that the first intramuscular hemorrhage was caused by abnormal coagulability. However, the coagulability at the time of the second hemorrhage was completely normal. We believe that the cause of the intramuscular hemorrhages observed in dermatomyositis patients with normal coagulability may imply the presence of intrinsic risk factors for spontaneous intramuscular hemorrhage in patients with dermatomyositis. Kissel et al. [6] found immune complex in the wall of intramuscular venules and arterioles, indicating that complement is deposited, bound, and activated within the intramuscular microvasculature in patients with dermatomyositis. In dermatomyositis, there is focal capillary depletion and the capillary density is significantly reduced [7]. These capillary changes were not revealed in

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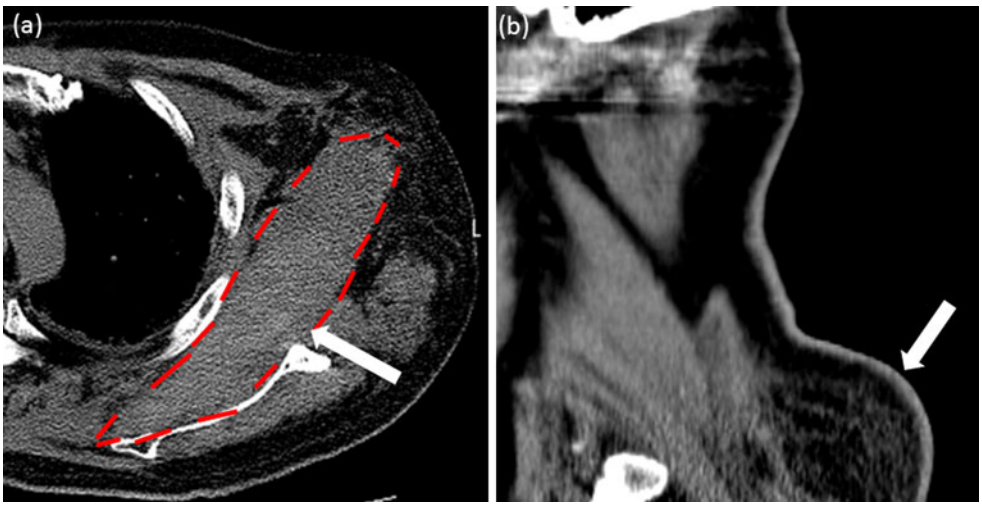

Fig. 1 HRCT scan of the back showing of **a** intramuscular hemorrhage from left deltoid (*arrow*), hematoma (enclosed by the *dotted line*) and **b** intramuscular hematoma in trapezius muscle (*arrow*). *HRCT* high-resolution computed tomography

Table 1 Muscle hematoma in dermatomyositis

Case	Age	Gender	Bleeding site	Heparin	Coagulability	Intravenous methylpredonisolone pulse	References
1	80	M	Left rectus sheath, oblique right thigh	Yes	APTT prolonged	Yes	[1]
2	50	F	Left rectus abdominis	No	Normal	No	[2]
3	11	F	Right reteroperitoneum	No	Normal	No	[2]
4	77	F	Left iliopsoas iliac, reteroperitoneum	Yes	APTT prolonged	Yes	[3]
5	64	F	Right reteroperitoneum, left rectus sheath	Yes	Normal	Yes	[4]
6	65	F	Iliopsoas both sides, thigh	Yes	APTT prolonged	Yes	[5]
7	60	M	Left trapezius	Yes	APTT prolonged	Yes	Our case

polymyositis. We suggest that the intramuscular hemorrhage in dermatomyositis was related to: (1) vessel-wall fragility caused by capillary vasculitis; (2) tissue fragility due to steroid treatment for dermatomyositis; and (3) prophylactic heparin.

We reported a case of dermatomyositis complicated with spontaneous intramuscular hemorrhage despite normal coagulability. Some other reports and our experience suggest that patients with dermatomyositis may have intrinsic risk factors for life-threatening intramuscular hemorrhage, and i.v. administration of UFH may affect the onset of hemorrhage.

References

1. Langguth DM, Wong RC, Archibald C et al (2004) Haemorrhagic myositis associated with prophylactic heparin use in dermatomyositis. Ann Rheum Dis 63:464–465
2. Orrell RW, Johnston HM, Gibson C et al (1998) Spontaneous abdominal hematoma in dermatomyositis. Muscle Nerve 21:1800–1803
3. Higashi Y, Mera K, Kanzaki T et al (2009) Fatal muscle haemorrhage attributable to heparin administration in a patient with dermatomyositis. Clin Exp Dermatol 34:448–449
4. Yamagishi M, Tajima S, Suetake A et al (2009) Dermatomyositis with hemorrhagic myositis. Rheumatol Int 29:1363–1366
5. Miwa Y, Muramatsu M, Takahashi R et al (2010) Dermatomyositis complicated with hemorrhagic shock of the iliopsoas muscle on both sides and the thigh muscle. Mod Rheumatol (Epub ahead of print)
6. Kissel JT, Mendell JR, Rammohan KW et al (1986) Microvascular deposition of complement membrane attack complex in dermatomyositis. N Engl J Med 314:329–334
7. Emslie-Smith AM, Engel AG (1990) Microvascular changes in early and advanced dermatomyositis: a quantitative study. Ann Neurol 27:343–56