Polymyositis is classified as an idiopathic inflammatory myopathy. It usually attacks adults and presents as progressively symmetric muscle weakness, especially of the proximal muscles [1]. Gastrointestinal involvement in adult dermatomyositis and polymyositis is usually mild. Recurrent abdominal pain and bloody diarrhea are clues of severe gastrointestinal involvement (for example, ulcers, ischemia or perforation) [2].

Spontaneous retroperitoneal hemorrhage is a lethal cause of acute abdomen that is most frequently related to drugs, coagulopathy and intra-abdominal tumors. In patients with polymyositis and dermatomyositis, acute abdomen is attributed to intestinal vasculitis causing ischemia, ulceration or perforation. Spontaneous retroperitoneal hemorrhage, however, has rarely been reported in patients with polymyositis. We report the case of a 65-year-old woman with newly diagnosed polymyositis and suspected thymoma who suffered from spontaneous retroperitoneal hemorrhage. She experienced two massive retroperitoneal hemorrhage episodes within 24 hours, which resulted in shock and required emergent angiographic embolization. There was no evidence of tumor, vasculitis or aneurysm from abdominal angiography and computed tomography.

Key Words: mediastinal tumor, polymyositis, spontaneous retroperitoneal hemorrhage (Kaohsiung J Med Sci 2008;24:436–40)

Case Presentation

A 65-year-old woman visited our rheumatology clinic in August 2005. Her medical history was unremarkable prior to this visit. She suffered from recurrent oral ulcers for 1 month without other skin or musculoskeletal symptoms. Laboratory examinations revealed an antinuclear antibody (ANA) titer of 1:640 and that the patient was negative for anti-dsDNA antibody. Early systemic lupus erythematosus was suspected at that time but not confirmed. She was treated with prednisolone at 10 mg/day, and the ulcer subsided within 1 week.

The patient remained asymptomatic until 7 months later, when progressive myalgia and muscle weakness in all four limbs occurred during regular daily exercise. Mild fever was also noted. She was referred by a primary medical physician to our emergency department for exacerbation of myalgia and muscle weakness. Initial laboratory examinations disclosed...
Spontaneous hemorrhage in a mediastinal tumor in a polymyositis patient

A creatine kinase level of 4,580 U/L (normal range, 22–261 U/L) and positive urine myoglobin.

On admission, myalgia progressed and proximal weakness of all four limbs of grade four (Medical Research Council grading) was found. Laboratory findings were as follows: ANA titer of 1:320 with a speckled pattern, negative for anti-Jo-1 antibody and anti-dsDNA antibody (ELISA, Pharmacia, Freiburg, Germany), and an erythrocyte sedimentation rate of 90 mm/hour. Electromyography revealed diffuse myopathy with active changes compatible with polymyositis. Pathologic examination of a muscle biopsy revealed inflammatory myopathy with lymphocytic infiltration. According to the aforementioned findings, we considered that this patient fulfilled the diagnostic criteria for polymyositis proposed by Bohan and Peter in 1975 [3].

An anterior mediastinal mass was discovered from chest X-ray at admission. Chest computed tomography (CT) disclosed a homogeneous anterior mediastinal mass without invasion to the surrounding soft tissue. There was no evidence of liver metastasis from abdominal sonography.

Hydrocortisone (300 mg/day) was administered, and significant improvement occurred within 1 week. Muscle power improved from grade four to grade five and creatine kinase level decreased from 5,808 U/L to 440 U/L. Surgery was scheduled but not carried out because of the following critical event.

On the 11th day of hospitalization, sudden abdominal pain occurred without any preceding traumatic event. Her blood pressure dropped to 86/54 mmHg. Nasogastric tube irrigation and anal digital examination showed no evidence of gastrointestinal bleeding. Paracentesis found fresh blood in the abdomen. Emergent abdominal CT showed retroperitoneal hematoma (Figure 1). Her white cell count was 8,760/μL, hemoglobin was 8.5 g/dL, platelet count was 384,000/μL, prothrombin time international normalized ratio was 1.05 and partial thromboplastin time was 23.6 seconds at that time. Emergent angiography revealed active bleeding from the superior duodenal-pancreatic artery (Figure 2A). Her vital status was stabilized after fluid resuscitation and angiographic embolization. Unfortunately, severe abdominal pain and refractory hypovolemic shock occurred 6 hours later. A second emergent angiography showed a new bleeding site in the superior mesenteric artery (Figure 2B). Embolization was performed again. The patient was later transferred to our intensive care unit. She died of sepsis 10 days later, without evidence of rebleeding on CT and laboratory data.

DISCUSSION

This patient fulfilled the full diagnostic criteria for definite polymyositis, including symmetric proximal muscle weakness, muscle biopsy evidence of myositis, increased levels of serum skeletal muscle enzymes and a characteristic electromyographic pattern [3].

Figure 1. Abdominal computed tomography shows retroperitoneal hematoma (arrow) collection between the aorta and superior mesentery artery and vein vasculature with associated continuous active bleeding and extravasation of contrast medium (thin arrow): (A) non-contrast; (B) contrast.
Anterior mediastinal tumors include thymoma, teratoma, seminoma, lymphoma, thyroid goiter and parathyroid adenoma [4]. The tumor was homogeneous without local invasion. Her peripheral blood cells, thyroid function, \( \alpha \)-fetoprotein and calcium level were normal. Considering the association with polymyositis and the above characters, thymoma was highly suspected in this case [4].

The patient had two massive retroperitoneal hemorrhage episodes. Spontaneous intra-abdominal hemorrhage is a rare condition. It was first described by Barber in 1909 [5] and could occur when there are ruptured aneurysms, tumors, coagulopathy, pregnancy, vasculitis and intra-abdominal cysts [6–9]. Long-term anabolic steroid abuse (nandrolone 30 times above the upper therapeutic dosage over 23 months) caused a huge subcapsular hepatic hematoma and subsequent intra-abdominal bleeding in a young male patient who used usual doses of mesterolone, clomiphene and chorionic gonadotropin at the same time [10]. To our knowledge, anterior mediastinal tumors have been associated with hemomediastinum caused by tumor hemorrhage, but not intra-abdominal hemorrhage [4]. Thymomas often cause thrombocytopenia, but not in this case [11]. Hence, the anterior mediastinal tumor in the patient was less likely to be the reason for the hemorrhage. Severe gastrointestinal complications in polymyositis patients usually result from vasculitis and are uncommon in adults [3]. Polymyositis and dermatomyositis induce small vessel disease, which is detectable by microscopy earlier than by angiography, and is even symptomatic [12,13]. T-cell-mediated cytotoxic reactivity against muscle fibers is detectable in polymyositis patients, whereas in dermatomyositis patients, vascular damage is caused by humoral mechanisms directed against small vessels and capillaries [12]. We presumed that the vessel disease caused by polymyositis in the form of acute spontaneous retroperitoneal vessel rupture happened before the lesion in our case was detectable by angiography.

Two cases with massive spontaneous intra-abdominal hemorrhage were reported, which were diagnosed as dermatomyositis and treated for many years [7]. Diffuse calcified vessels were deemed to be evidence of severe long-term inflammation. Fragile vessels or chronic steroid administration was presumed to be the reason for hemorrhage [7]. However, our patient had different characteristics compared with those in previous reports, being a newly diagnosed case and having received only an 8-day course of steroid therapy. No significant associated drug or medical history was identified. Angiography showed no evidence of vasculitis, aneurysm, arterial occlusion or calcification of intra-abdominal vessels.

Figure 2. (A) Selective gastroduodenal arteriography reveals superior duodenopancreatic artery active bleeding (arrow) with contrast medium pooling into the retroperitoneum, instead of characteristic angiography imaging evidence of vasculitis, local calcification and aneurysm formation. (B) The second angiography shows a new bleeding site along the jejunum branch of the superior mesentery artery (arrow).
Retroperitoneal bleeding is a lethal problem that warrants early detection for better prognosis [7]. We present a case of retroperitoneal bleeding and suggest a possible cause for acute abdomen in this polymyositis patient.

REFERENCES

縱膈腔腫瘤及多發性肌炎併發自發性後腹腔出血
— 一個案報告

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自發性後腹腔出血是急性腹痛症中可能造成死亡的疾病之一，大多與藥物、凝血功能不佳或腹內腫瘤有關。在多發性肌炎和皮肌炎的病人中，急性腹痛症多是因為腸血管炎造成缺血、潰瘍甚至穿孔而引起。多發性肌炎病人合併有自發性後腹腔出血，是幾乎沒有被報導過的案例。我們的病患是一位 65 歲女性，剛被診斷為多發性肌炎及疑胸腺瘤，發生自發性後腹腔出血。她在 24 小時內有兩次大量後腹腔出血，引致休克，必須兩次緊急血管栓塞術方能止血。在血管攝影及電腦斷層中，並沒有發現病人有腹內腫瘤、血管炎或血管瘤。

關鍵詞：縱膈腔腫瘤，多發性肌炎，自發性後腹腔出血
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