Life-threatening spontaneous hemothorax in a patient with thrombotic thrombocytopenic purpura

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Abstract
Spontaneous hemothorax in the left pleural space occurred suddenly in a patient with thrombotic thrombocytopenic purpura (TTP). In spite of massive blood transfusion, the hemorrhage could not be stopped. The patient suffered shock due to tension hemothorax and hypovolemia, resulting in cardiac arrest. After successful cardiopulmonary resuscitation, surgical hemostasis was performed. The main cause of the bleeding was rupture of the left intercostal vein. TTP is a severe microvascular occlusive thrombotic microangiopathy that can induce congestion, vasculitis, and ischemia. This mechanism is thought to have been involved in the rupture of the intercostal vein in the present patient.

Key words Thrombotic thrombocytopenic purpura · Hemothorax · Cardiac arrest

Introduction
Thrombotic thrombocytopenic purpura (TTP) is a severe microvascular occlusive disorder characterized by severe thrombocytopenia, fragmented erythrocytes with hemolysis, acute renal failure, fever, and neurological disturbance [1]. TTP is a potentially fatal disease, and patients can develop acute respiratory distress syndrome [2], intrapulmonary hemorrhage [3], myocardial ischemia [4, 5], and life-threatening arrhythmia [6]. Spontaneous hemothorax is an extremely unusual event. Although a few cases have been reported in patients with hematological diseases [7–10], to our best knowledge there has been no case of spontaneous hemothorax in patients with TTP. The following is the first case report of TTP in which massive hemorrhage due to rupture of an intercostal vein resulted in life-threatening tension hemothorax.

Case report
A 59-year-old man was referred to our hospital for management of chronic renal failure (CRF) by hemodialysis. He had been hospitalized at another hospital for examination of proteinuria and renal dysfunction 2 months previously, and other typical symptoms of TTP, including hemolytic anemia, thrombocytopenia, delirium, and confusion had developed within 5 hospital days. A diagnosis of TTP had been made by a hematologist, according to laboratory data showing von Willebrand factor-cleaving metalloprotease (ADAMTS13) deficiency [1, 11]. The patient had undergone 14 plasma exchanges during 17 days, and thereby his neurological symptoms and platelet count had been improved. However, his renal function had not recovered, resulting in CRF.

On admission to our hospital, laboratory data showed mild anemia and thrombocytopenia (hemoglobin, Hb, 10.6 g·dl⁻¹; platelets, 82,000·µl⁻¹). He underwent hemodialysis 3 times per week. On the tenth hospital day, he suddenly complained of left back pain and mild dyspnea in the morning. His respiratory difficulty gradually deteriorated over the next 8 h. A chest roentgenogram in the evening showed left pleural effusion. Aspiration of the left pleural cavity revealed bloody effusion, and approximately 750 ml of blood was removed; however, the dyspnea progressively got worse. About 90 min after the aspiration procedure, cardiovascular collapse occurred. Although a rapid infusion of 5% albumin and crystalloid solution was administered, systolic blood pressure decreased to below 80 mmHg. A chest roentgenogram showed marked left pleural effusion and mediastinal shift to the right (Fig. 1). He became comatose and his pupils became dilated. Immediately, orotracheal intubation was performed to initiate intermittent positive-pressure ventilation with 100% oxygen. A 20-Fr trocar catheter was inserted into the left pleural cavity from the fifth intercostal space, which was
continuously aspirated at −20 cmH2O. Arterial blood gas analysis revealed severe metabolic acidosis (pH, 7.065; base excess [BE], −16.9 mEq·l−1; PaCO2, 41.6 mmHg; PaO2, 339.5 mmHg) caused by hemorrhagic shock. Trans-fusions of packed blood cells (PBC) and fresh frozen plasma (FFP) were started via an 11-Fr central venous catheter; however, the pleural bleeding could not be stopped. The total amount of bleeding from the left pleural cavity exceeded 3100 ml. Cardiovascular stability was not achieved even after massive transfusion (PBC, 10 U and FFP, 15 U). Blood pressure and heart rate were dynamically fluctuating; we decided on emergency thoracotomy for surgical hemostasis.

The patient was transferred to the operating room with a continuous intravenous infusion of noradrenaline (1 μg·kg−1·min−1) and intermittent positive-pressure ventilation. On arrival, the patient was in a comatose state (Glasgow coma scale 3), and electrocardiogram and direct arterial pressure via the right femoral artery showed marked hypotension (40/20 mmHg) and bradycardia (20 bpm). Although rapid blood transfusion and intravenous vasopressin 20-μg administration were performed, cardiac asystole soon occurred. Cardiopulmonary resuscitation (CPR) with chest compression and intravenous administration of epinephrine (total dose, 12 mg) and vasopressin (total dose, 40 μg) was started immediately. Approximately 5 min later, spontaneous cardiac rhythm appeared again. Then the tracheal tube was exchanged for a 37-Fr double-lumen tube for one-lung ventilation. Anesthesia was maintained with continuous infusion of propofol, intermittent bolus of fentanyl, and vecuronium with titration. After a left thoracotomy had been performed, massive blood and clot (total, 3658 g) were aspirated from the left pleural space. The trocar catheter was found to be occluded by a clot. The main cause of bleeding was rupture of the left intercostal vein near the eighth rib and vertebra. Hemostasis at this point was achieved by ligation of the ruptured vessel and compression with a Dacron felt patch; his cardiovascular state was then markedly improved. We also found several points of oozing surrounded by inflammatory tissue; however, complete hemostasis of these areas was difficult. His platelet count had decreased by 20 000 · μl−1 at this time, and this was also likely to be involved in the oozing.

After the operation, continuous positive-pressure ventilation was continued. Although the pleural bleeding continued until the second postoperative day, his cardiovascular state was stable. The morning after the operation, his consciousness had recovered and no neurological deficit was observed. Successful weaning from mechanical ventilation was achieved 2 weeks after the operation. However, typical symptoms of TTP, including hemolytic anemia, jaundice, and neurological deficit emerged again beginning from 1 month after the operation. The patient underwent six plasma exchanges; however, he died of cardiorespiratory failure 2 months after the operation.

Discussion

Hemothorax is usually caused by chest trauma, malignancy, pulmonary embolism, aortic aneurysm, and infectious disease. Spontaneous hemothorax due to hematological disease is a rare event. Although there have been a few reported cases of spontaneous hemothorax in patients with idiopathic thrombocytopenic purpura (ITP) [9, 10], the pathogenesis of ITP is different from that of TTP. The main pathological feature of TTP is a severe microvascular occlusive thrombotic microangiopathy, which is caused by the activation of platelet aggregation and the adhesion of platelets to capillary endothelial walls due to the accumulation of von Willebrand factor multimers [1]. In TTP, organ hypoxemia caused by microthrombi manifests as several characteristic symptoms, including neurological abnormalities and renal failure. The platelet count is not always at a critical level to cause spontaneous bleeding as was shown in the present patient (82 000 · μl−1). Coagulation test results are usually normal unless ischemic tissue damage causes secondary disseminated intravascular coagulation [12]. Martinez et al. [3] reported a patient with intrapulmonary hemorrhage seen as the primary manifestation of TTP. Postmortem examina-