인공 슬관절 전치환술 후 발생한 우심부전을 동반한 혈전성혈소판감소자반증

¹대구가톨릭대학교 의과대학 내과학교실, ²울산대학교 의과대학 서울아산병원 호흡기내과 및 중환자의학과

심상우¹ · 임채만² · 고윤석² · 홍상범²

Acute Thrombotic Thrombocytopenic Purpura with Right Heart Failure Following Total Knee Replacement Surgery

Sangwoo Shim¹, Chae-Man Lim², Younsuck Koh², and Sang-Bum Hong²

¹Department of Internal Medicine, Catholic University of Daegu School of Medicine, Daegu; ²Department of Pulmonary and Critical Care Medicine, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea

Thrombotic thrombocytopenic purpura (TTP) is a life-threatening condition characterized by microangiopathic hemolytic anemia, thrombocytopenia, renal insufficiency, neurological abnormalities, and fever. Cardiac involvement is not uncommon and can be fatal; however, right ventricular heart involvement after surgery is rare. Here, we report a case of TTP presenting with right ventricular heart failure after total knee replacement surgery. TTP was successfully treated with four rounds of plasma exchange. The patient made a full recovery and was discharged after 11 weeks. (Korean J Med 2018;93:220-223)

Keywords: Purpura, Thrombotic thrombocytopenic; Orthopedics; Heart failure

INTRODUCTION

Thrombotic thrombocytopenic purpura (TTP) is a lethal condition characterized by microangiopathic hemolytic anemia (MAHA), thrombocytopenia, renal insufficiency, neurological abnormalities, and fever [1]. The incidence of acute heart failure associated with TTP is reported to be 9.5% and is considered a poor prognostic factor [2]. The vast majority of heart failures in patients with TTP are associated with the left side of the heart, with few reports of right ventricular heart failure (RVHF). Here, we present a case of TTP with RVHF following total knee replacement (TKR) surgery.

CASE REPORT

An 80-year-old woman presented at our emergency depart-

Correspondence to Sang-Bum Hong, M.D.

Tel: +82-2-3010-3893, Fax: +82-2-3010-6968, E-mail: sbhong@amc.seoul.kr

Copyright © 2018 The Korean Association of Internal Medicine

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (http://creativecommons.org/licenses/by-nc/3.0/) which permits unrestricted noncommercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

Received: 2016. 8. 22

Revised: 2017. 10. 16

Accepted: 2017. 10. 29

Department of Pulmonary and Critical Care Medicine, Asan Medical Center, University of Ulsan College of Medicine, 88 Olympic-ro 43-gil, Songpa-gu, Seoul 05505, Korea

ment in a stuporous condition. The patient had undergone TKR at a different hospital on the previous day but began exhibiting significant changes in mental status within 24 hours after the surgery. The patient had no history of cardiac disease, but she did suffer from hypertension. The patient had been taking atenolol, felodipine, and domperidone prior to the operation. Examination in the emergency department revealed the following: blood pressure, 65/39 mmHg; pulse rate, 98 beats per minute; respiratory rate, 30 breaths per minute; and body temperature, 35.0°C. Arterial blood gas analysis performed while she was breathing oxygen (15 L/min) through a reservoir mask revealed the following: pH, 7.350; PaCO₂, 33.0 mmHg; PaO₂, 48.0 mmHg; HCO₃, 18.0 mmol/L; and O₂ saturation, 83.4%. Crackles were heard in both lung fields. The wound of left knee was clear, with no erythema, swelling, discharge, bleeding, or dehiscence. She underwent endotracheal intubation, was placed on mechanical ventilation, and was treated with a large volume of crystalloid, high-dose vasopressors, and inotropics. Initial laboratory findings revealed a white blood cell count of 17,600/mm³, a hemoglobin level of 15.0 g/dL, and a platelet count of 128,000/mm3. Blood coagulation tests (prothrombin time [PT] and activated partial prothrombin time [aPTT]) were



Figure 1. Chest radiography on the day of admission shows cardiomegaly and pulmonary edema.

normal. The blood urea nitrogen level was 34 mg/dL and the serum level of creatinine was 2.0 mg/mL. The levels of lactate dehydrogenase and total bilirubin were 447 IU/L and 2.1 mg/dL, respectively. Cardiac enzymes were mildly elevated: CK-MB, 8.7 g/mL, and troponin-I, 0.637 µg/mL. Brain levels of natriuretic peptides were 1,444 pg/mL and the lactate level was 4.0 mmol/L. Chest radiography indicated bilateral pulmonary edema (Fig. 1). Electrocardiogram results were normal (Fig. 2). Echocardiography revealed normal left ventricular function; however, the right ventricle was dilated and showed reduced contractility. Left ventricular ejection fraction was 56%, with left ventricular dimensions of 33 mm at diastole and 23 mm at systole; the left atrial dimension was 19 mm. The estimated pulmonary artery systolic pressure was 39 mmHg (Fig. 3); however, dynamic chest computed tomographic angiography showed no evidence of pulmonary embolism (Fig. 4). The patient was in an anuric state from the first day of admission. On day 3 of hospital admission, hemoglobin levels fell to 8.6 g/dL and the platelet count fell to 20,000/mm³, with no associated increases in PT or aPTT. No microorganisms were detected in cultures from blood, urine, sputum, and wound. A peripheral blood smear contained schistocytes. Based on these results, the patient was diagnosed with postoperative TTP following TKR surgery, with evidence of MAHA, thrombocytopenia, renal failure, and altered mental status. The patient underwent four rounds of plasma exchange, after which she showed no signs of TTP. She was discharged after 11 weeks.



Figure 2. An electrocardiogram on the day of admission shows normal sinus rhythm and no specific abnormalities.

- 대한내과학회지: 제 93 권 제 2 호 통권 제 681 호 2018 -



Figure. 3. A transthoracic echocardiogram on the day of admission shows a dilated right ventricle and reduced right ventricular contractility; the function of the left ventricle is normal.



Figure. 4. A Chest computed tomographic angiogram on day 3 of hospitalization shows no evidence of pulmonary embolism and right dominant bilateral pleural effusion with passive atelectasis.

DISCUSSION

TTP, initially described by Moschcowitz in 1924, is a life-threatening disease [1]. The mechanism underlying acquired TTP involves the formation of unusually large multimers of von Willebrand factor due to a severe reduction in the level of disintegrin and metalloproteinase with a thrombospondin type 1 motif, member 13 (ADAMTS13) [3]. In most patients with acquired TTP, the level of plasma ADAMTS13 activity is < 5% of normal values [4]. We did not confirm the level of

ADAMTS13 activity in the patient reported herein, which could be seen as a limitation; however, the patient showed many of the classic features of TTP, which makes a misdiagnosis unlikely.

The development of TTP after orthopedic surgery is not common [5-7]. Among cases of TTP that do develop after orthopedic surgery, the case reported here is unique because it was accompanied by RVHF. The reported incidence of cardiac involvement in TTP is 9.5%, with a mortality rate of 38% [2]. However, only left ventricular heart failure has been reported. The patient described herein had RVHF, with normal left ventricular systolic function. Treatment of TTP resulted in an improvement in right ventricular function, which suggests that RVHF may be associated with TTP, particularly after TKR surgery.

As the initial presentation was dyspnea due to pulmonary edema, there is a possibility of transient left ventricular dysfunction associated with stress-induced cardiomyopathy. In cases of right ventricular involvement in stress-induced cardiomyopathy, these symptoms are typically accompanied by lower left ventricular ejection fraction and higher incidence of pulmonary edema [8]. Here, we observed only right ventricular dysfunction at the time of admission, with insufficient time for left ventricular function to have recovered if a bilateral failure were to have occurred. Alternatively, cardiac involvement of TTP has been shown to manifest as arrhythmia in 15% of cases [9], which suggests that a brief arrhythmia may have caused elevated left atrial end-diastolic pressure to induce pulmonary edema.

In this patient, we treated TTP using plasma exchange. Previous studies have recommended daily plasma exchange in cases of acquired acute idiopathic TTP [10]. Plasma exchange removes von Willebrand factor multimers and autoantibodies against ADAMTS13 and provides additional metalloprotease enzymes. In this case, plasma exchange led to a full recovery, which indicates that plasma exchange is a useful therapy for patients with TTP involving right ventricular failure, as well as for those with more traditional TTP symptoms. Taken together, the data presented here suggest that while RVHF is a rare complication in patients with TTP, the condition can be treated effectively with plasma exchange.

중심 단어: 혈전성 혈소판감소자반증; 정형외과; 심부전

REFERENCES

- Moschcowitz E. Hyaline thrombosis of the termial arterioles and capillaries: a hitheto undescribed disease. Proc NY Pathol Soc 1924;24:21-24.
- Gami AS, Hayman SR, Grande JP, Garovic VD. Incidence and prognosis of acute heart failure in the thrombotic microangiopathies. Am J Med 2005;118:544-547.
- Crowther MA, George JN. Thrombotic thrombocytopenic purpura: 2008 update. Cleve Clin J Med 2008;75:369-375.
- Tsai HM, Rice L, Sarode R, Chow TW, Moake JL. Antibody inhibitors to von Willebrand factor metalloproteinase and increased binding of von Willebrand factor to platelets in ticlopidine-associated thrombotic thrombocytopenic purpura. Ann Intern Med 2000;132:794-799.
- Kathula SK, Kamana M, Naqvi T, Gupta S, Chang JC. Acute thrombotic thrombocytopenic purpura following orthopedic surgery. J Clin Apher 2002;17:133-134.
- Iosifidis MI, Ntavlis M, Giannoulis I, Malioufas L, Ioannou A, Giantsis G. Acute thrombotic thrombocytopenic purpura following orthopedic surgery: a case report. Arch Orthop Trauma Surg 2006;126:335-338.
- Yenigun EC, Bardak S, Piskinpasa SV, et al. Acute thrombotic thrombocytopenic purpura following orthopedic surgery: case report and review of the literature. Ren Fail 2012;34:937-939.
- Haghi D, Athanasiadis A, Papavassiliu T, et al. Right ventricular involvement in Takotsubo cardiomyopathy. Eur Heart J 2006;27:2433-2439.
- Hawkins BM, Abu-Fadel M, Vesely SK, George JN. Clinical cardiac involvement in thrombotic thrombocytopenic purpura: a systematic review. Transfusion 2008;48: 382-392.
- Rock GA, Shumak KH, Buskard NA, et al. Comparison of plasma exchange with plasma infusion in the treatment of thrombotic thrombocytopenic purpura. N Engl J Med 1991;325:393-397.