CASE REPORT

Acute Pulmonary Embolism due to Paget-Schroetter Syndrome

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Abstract

A 17-year-old Japanese male athlete presented to the emergency department at our hospital with a chief complaint of exertional dyspnea. Although there were no significant findings in the right and left upper extremities on a physical examination, a chest computed tomography scan showed bilateral multiple thrombosis in the pulmonary arteries, indicating pulmonary thromboembolism, and deep vein thrombosis in the left subclavian vein. Upper limb venography showed interruption of the left subclavian vein (so-called Paget-Schroetter syndrome; PSS). We herein report this rare case of PSS that led to pulmonary thromboembolism in a young, male field athlete.

Key words: Paget-Schroetter syndrome, deep vein thrombosis, pulmonary thromboembolism

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Introduction

Pulmonary thromboembolism is commonly caused by deep vein thrombosis (DVT) in the lower limbs, although it rarely occurs in the upper limbs (1, 2). Paget-Schroetter syndrome (PSS) has been reported to be a rare presentation of primary axillary subclavian vein thrombosis with underlying thoracic outlet syndrome following prolonged upper extremity exertion, that typically occurs in young men (3, 4). The primary complication of PSS is post-thrombotic syndrome due to pulmonary artery embolism. In this report, we describe a rare case of PSS that led to pulmonary thromboembolism in a young, male field athlete.

Case Report

A 17-year-old adolescent, who was a track and field athlete, was admitted to our emergency department with progressive exertional dyspnea that had persisted for the previous seven days. His consciousness was clear. His height was 168 cm and his weight was 65 kg. A physical examination revealed a temperature of 37.0°C, pulse rate of 120/min, respiratory rate of 23/min, and blood pressure of 120/80 mmHg with no external injuries. There was no swelling, edema, cyanosis, or laterality in the face or upper and lower extremities. Although the second heart sound was slightly split, no murmurs or crackles were detected. A blood gas analysis showed hypoxia, with a pH of 7.45, partial pressure of arterial oxygen (PaO₂) of 58 Torr, and partial pressure of carbon dioxide in arterial blood (PaCO₂) of 36 Torr on room air. Coagulation tests revealed increased levels of fibrin/fibrinogen degradation products at 35.0 μg/mL (normal: ≤5 μg/mL) and D-dimer at 16.5 μg/mL (normal: ≤1.0 μg/mL). The activities of protein C (95%), protein S (104%), and antithrombin III (105%), and the levels of thrombomodulin (2.2 FU/mL), and homocysteine (14.1 mmol/mL) were normal. There were no collagen disease-related autoantibodies or, antiphospholipid antibodies. A chest X-ray examination showed normal findings (Fig. 1), while electrocardiography demonstrated incomplete right bundle branch block (Fig. 2). Cardiac ultrasonography revealed dilation of the right atrium and ventricle, and deviation of the septal wall toward the left ventricle, indicating the presence of pulmonary hypertension (estimated systolic pulmonary arterial pressure; 65 mmHg; Fig. 3). A chest CT scan showed multiple thrombi in the bilateral pulmonary arteries (Fig. 4). Although there were no signs of DVT in the lower limbs, DVT was de-
The pulmonary thromboembolism was treated with tissue plasminogen activator (tPA, monteplase, 27,500 IU/kg), and heparin (activated partial thromboplastin time 1.5 to 2 times), followed by warfarin (prothrombin time-international normalized ratio between 2.0 and 3.0). The patient’s dyspnea immediately resolved after tPA and heparin treatment. Two weeks after the administration of anticoagulant therapy, upper limb venography (Fig. 5) showed obstruction of the left subclavian vein with a collateral circulation in the cervical region. There were no other abnormalities in the superior vena cava or right subclavian vein. A follow-up CT scan, performed one week after the initial therapy, showed that the bilateral thrombi in the pulmonary arteries had decreased, and that the left costoclavicular space was narrower than the right costoclavicular space, indicating that the left subclavian vein was interrupted (Fig. 6). We considered that the patient had left subclavian vein thrombosis and obstruction, caused by PSS. The elements of the chest wall, including the clavicle, showed no abnormalities. Furthermore, the numerous collaterals observed on venography suggested that the venous obstruction was chronic. Therefore, we did not administer surgical or interventional therapy, including intravenous metallic stent insertion or balloon venoplasty. One year after the administration of anticoagulant therapy, a chest CT scan showed that the bilateral thrombi in the pulmonary arteries...
Discussion

Pulmonary thromboembolism is caused by DVT, originating from the lower limbs in over 90% of cases, and rarely from the upper limbs (5). PSS results from venous endothelial damage due to overwork of the upper limbs, which typically occurs in young healthy athletes (6). The mechanisms underlying this type of thrombosis in PSS patients are considered to be as follows: the costoclavicular space narrows when the shoulder moves posteroinferiorly or when the arm is excessively rotated externally, whereby the subclavian vein is compressed between the first rib and the clavicle (7, 8). Actions that repeatedly and excessively cause such compression can lead to the onset of phlebitis and DVT (7, 8). Eventually, the pulmonary thromboembolism progresses to cause symptoms of exertional dyspnea and syncope. Idiopathic subclavian vein thrombosis is usually accompanied by edema, cyanosis, and pain in the ipsilateral upper extremity; however, our patient had no such obstructive symptoms, probably because the venous obstruction was chronic.

As previously indicated, PSS should be considered a serious condition, as it may result in severe sequelae and permanent disability, if not promptly and properly treated (9, 10). The treatment of PSS usually involves thrombolytic therapy, percutaneous venoplasty, and surgical decompression of the thoracic outlet (11, 12). In the clinical setting, the indications for surgical intervention have been debated; i.e., whether to perform surgical therapy only in subjects with persistent or recurrent symptoms even after thrombolysis, or to provide early decompression in all patients (13). Although thrombectomy should be performed within 48 hours of the onset of obstruction, if possible, it is
difficult to remove the thrombi after they exhibit strong adherence to the vessel wall. Resection of the first rib, the intravenous insertion of metallic stents, and balloon venoplasty are other therapeutic options; however, these procedures remain controversial. In the present case of a chronic occluded subclavian vein, we administered only anticoagulant therapy with warfarin to control the prothrombin time-international normalized ratio between 2.0 and 3.0. If the chronic obstruction in the left subclavian vein remains stable, the anticoagulation may be discontinued in the future after careful observation.

As presented in this case, the possibility of PSS should be considered in patients with pulmonary thromboembolism, especially young athletes.

The authors state that they have no Conflict of Interest (COI).

**References**

4. Spencer TR, Lagace RE, Waterman G. Effort thrombosis (paget-