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Case Report

Paget–Schroetter syndrome accompanied by pulmonary thromboembolism: A case report

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ABSTRACT

A 17-year-old male, who was involved in a baseball club, presented to our emergency department with the complaint of gradual onset of swelling of his right arm. Contrast-enhanced computed tomography showed obstruction of the proximal portion of the right subclavian vein and pulmonary thromboembolism. Venography confirmed an occluded right subclavian vein. The patient was diagnosed with right subclavian vein thrombosis, which is referred to as Paget–Schroetter syndrome (PSS). An ultrasonography for the affected subclavian vein was helpful not only for making an accurate diagnosis of PSS, but also for verifying dynamic venous flow changes depending on the forearm position.
<**Learning Objective:** This paper aims to describe the usefulness of ultrasonography. A 17-year-old male was diagnosed with Paget–Schroetter syndrome (PSS). In this case, we found ultrasonography is important for verifying dynamic venous flow changes depending on the forearm position. Holding the forearm in a pronated and downward position may have been critical for maintaining good venous flow with prompt resolution of symptoms. Ultrasonography may play a crucial role in assessing an appropriate treatment strategy for PSS.>

Introduction

Paget–Schroetter syndrome (PSS) is an unusual cause of upper extremity deep venous thrombosis (DVT). DVT refers to subclavian vein thrombosis associated with strenuous and repetitive activities of the upper extremities. PSS accounts for 10–20% of all arm DVT [1,2] and is also described as “effort thrombosis”. Thoracic outlet abnormalities and repetitive arm movements, such as a pitching arm, causing vascular injuries, may be common predisposing factors for venous thrombosis [1,2]. We report a rare case of PSS complicated by pulmonary embolism.

Case report

A 17-year-old male, a high-school student, presented to our emergency department with the complaint of gradual onset of redness and swelling of the right upper extremity. His past medical history was unremarkable. He had no history of arm and shoulder trauma and no family history of thrombosis. He joined a baseball team as a right-handed pitcher. He spent a lot of time practicing hard.

On admission, his general status was unremarkable. His height was 175 cm and weight was 67 kg. A physical examination showed a temperature of 36.2 °C, pulse rate of 70 beats/min, respiratory rate of 16 breaths/min, blood pressure of 128/70 mmHg, and the oxygen saturation was 96% in room air. Heart and respiratory sounds were clear. His right forearm showed remarkable swelling up to his shoulder and its movement was severely restricted. He was forced to keep his arm in non-stretched pronated position. A chest X-ray examination and electrocardiography were within the normal range. Laboratory tests indicated mild inflammatory change, with a white blood cell count ($7.8 \times 10^3/\mu L$) and C-reactive protein (1.04 mg/dL). On coagulation tests, D-dimer was slightly increased (2.67 μg/ml: normal range 0.22–0.50 μg/ml), but prothrombin and partial thromboplastin times were normal. The level of anticoagulation factors, such as protein C, protein S, and AT-III activity were also within normal ranges. Antinuclear

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antibody, anti-cardiolipin antibody, and lupus anticoagulant were negative.

Contrast-enhanced computed tomography (CT) showed multiple thromboses in the pulmonary artery (Fig. 1A). Although there were no signs of DVT in the lower limbs, the right proximal subclavian vein was occluded with thrombus formation on the image (Fig. 1B). Lung perfusion scintigraphy showed multiple segmental perfusion defects with normal ventilation images, which was compatible with multiple pulmonary thromboembolisms (Fig. 1C). Venography confirmed obstruction of the proximal portion of the right subclavian vein with collateral venous flow toward the superior vena cava (Fig. 1D). We diagnosed him with right upper extremity DVT accompanied by pulmonary thrombosis, also known as PSS. Cardiac sonography showed dilation of the right ventricle and moderate pulmonary hypertension. An ultrasonography of the right subclavian vein detected venous thrombosis in the proximal site of the subclavian vein as shown by CT. When pulling down and/or internally rotating his right arm, forward flow of the subclavian vein was obviously detected by color flow mapping (Fig. 2A). Venous blood flow was abruptly interrupted when lifting up and/or externally rotating (Fig. 2B). Although CT images showed complete occlusion of the subclavian vein, ultrasonography clearly demonstrated that venous flow was intermittently revascularized depending on the arm position. This provided useful information to treat his swollen right arm. Keeping his right arm pulled down and internal rotation was important for maintaining good venous flow. After admission, he was treated with a urokinase-type plasminogen activator (administered 240,000U/day from 2nd to 4th day, 120,000U/day from 5th to 7th day, and 60,000U from 8th to 10th day) and unfractionated heparin through his right cephalic vein for 1 week, followed by rivaroxaban 30 mg/day (Fig. 3). After several days on anticoagulation and thrombolysis therapy, he became asymptomatic. He completely recovered and multiple thrombi in the pulmonary artery totally disappeared. On the 19th day, he was uneventfully discharged.

Discussion

An unusual cause of subclavian vein thrombosis was described by Paget and von Schroetter [3,4], and this was called PSS. Thoracic outlet abnormalities and/or repetitive movements causing vascular injuries are well known as common predisposing factors for PSS. [5,6] In this article, we report a case of PSS in a young baseball player, complicated by pulmonary thromboembolism.

The thoracic outlet region contains the brachial plexus, the subclavian artery, and the subclavian vein. The subclavian vein is located at the front of the anterior scalene muscle, so that the subclavian vein may facilitate mechanical compression between the first rib and the clavicle. Repetitive movements of the forearm may lead to intimal injury of the subclavian vein due to mechanical stimuli and may be prone to causing thrombus formation [7,8]. As the patient had exercised pitching hard his right arm muscle heavily developed hypertrophy of the anterior scalene and subclavius muscles that may have led to compression of the vessel into the first rib that may contribute to the cause of PSS [9].

As is the case with lower extremity DVT, we administered thrombolytic agent and anticoagulant drug. After venous thrombus was successfully lysed, venous flow ran mainly from subclavian vein (not collateral veins) to the central vein, while organic stenosis caused by the muscle hypertrophy remained. Venous flow pattern examined by ultrasonography was dramatically changed depending on the patient's forearm positions (Fig. 2). An externally supinated and/or upward position of the affected arm caused subclavian venous flow to be abruptly interrupted, while an internally pronated and downward position led to good

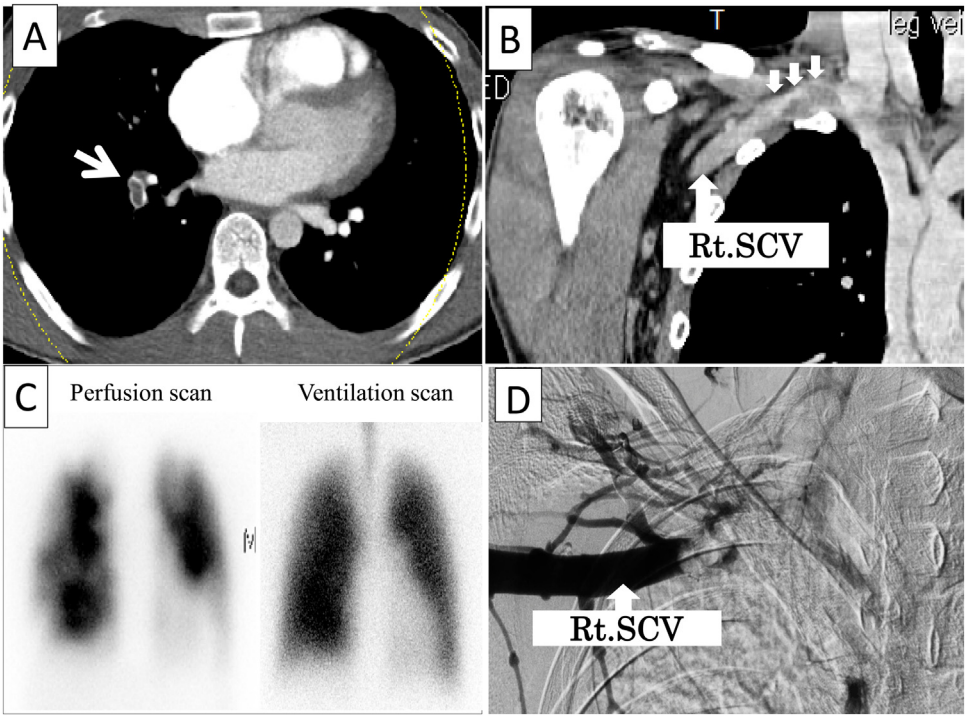

Fig. 1. (A) Chest computed tomography (CT) scan showing bilateral multiple thrombi in the pulmonary arteries. (B) Chest contrast enhanced CT scan showing a filling defect in the right subclavian vein (SCV) and the compressed vein at the thoracic inlet portion. (C) Lung perfusion scintigraphy showing multiple segmental perfusion defects in a wedge-shaped configuration with normal ventilation images. (D) Venography showing the obstructed right subclavian vein and the collateral circulation toward superior vena cava.

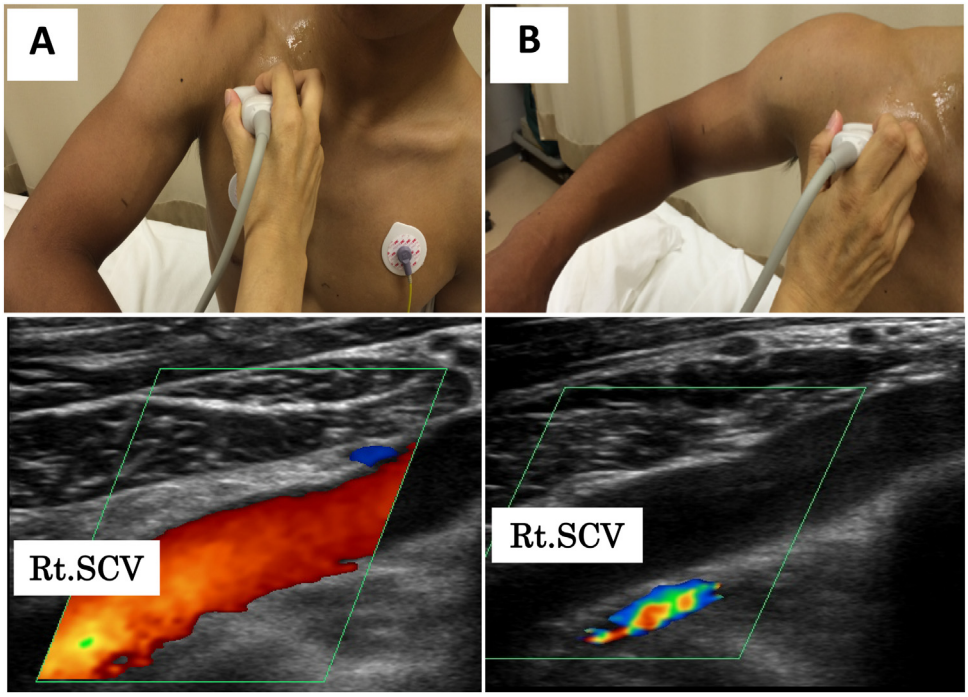

Fig. 2. Positional relationship of the patient arm and the right subclavian vein (Rt. SCV) flow. (A) When pulling down and/or internally rotating his right arm, the SCV forward flow was obviously detected by color flow mapping. (B) When lifting up and externally rotating, the flow was interrupted.

venous flow. These physical findings were repeatedly confirmed by ultrasonography.

Although CT angiography and subclavian venography were of diagnostic value for upper extremity DVT, we did not observe the unexpected observations of on-and-off venous flow disturbance depending on the arm position without performing ultrasonography. In patients who are suspected of upper extremity DVT, an ultrasonography is important for confirming an accurate diagnosis of PSS and verifying dynamic venous flow changes, depending on the forearm position. In our case, holding the forearm in a pronated and downward position may have been critical for maintaining good venous flow with prompt resolution of symptoms. Ultrasonography has several potential advantages, such as being noninvasive and easily repeated and allowing the evaluation of the deep and superficial venous systems [10]. This phenomenon could have been verified by other modalities such as venography and magnetic resonance imaging venography, although we did not perform these in this case.

Other treatments for PSS are still under clinical investigation, such as new thrombolysis agents, percutaneous thrombectomy,

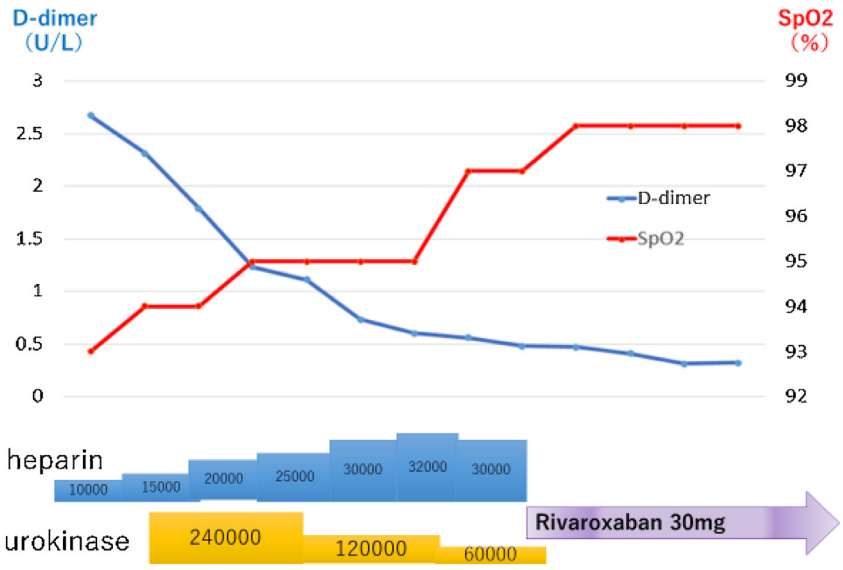

Fig. 3. Progress after admission. After admission, he was treated with urokinase-type plasminogen activator and unfractionated heparin through his right cephalic vein for a week, followed by rivaroxaban 30 mg/day (direct oral factor Xa inhibitor).

and surgical venoplasty. Catheter-directed thrombolysis may be considered in patients with DVT of an upper extremity who present with extensive swelling and functional impairment of the arm [9]. Percutaneous mechanical catheter interventions may also be considered in patients who have persistent, severe symptoms of DVT when anticoagulant and thrombolysis therapies are unsuccessful [9]. Aggressive treatment, such as the "first rib resection", may be approved when critical thrombus remains at the thoracic outlet site. Whether these treatments have sufficient long-term patency remains controversial, although invasive surgical procedures for DVT have been reported to have a higher rate of venous patency in some previous studies [9,10]. Conservative anticoagulation therapy is still considered as a standard therapy for effort thrombosis, such as PSS. Subsequent long-term data are needed to assess whether ultrasonography based on long-term medical management may be helpful to reduce residual symptoms, disability, and recurrent thrombus formation.

Conclusions

We experienced a case of PSS, known as effort thrombosis, accompanied by pulmonary thromboembolism. Thoracic outlet abnormalities and repetitive forearm movements may be predisposing factors for PSS. An ultrasonic examination of the affected subclavian vein was helpful not only for making an accurate diagnosis of PSS, but also for verifying dynamic venous flow changes depending on the arm positions. Ultrasonography may play a crucial role in assessing an appropriate treatment strategy for PSS.

Conflict of interest

The authors state that they have no conflict of interest (COI).

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