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Case Report

Two Surgical Patients with Paget–Schroetter Syndrome Presenting with Atypical Pathophysiology: Non-Effort Thrombosis

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Paget–Schroetter syndrome (PSS) is an upper extremity thrombosis occurring in the axillary and subclavian veins. PSS is also known as "effort thrombosis," because it is usually associated with repetitive and strenuous activities of the upper limbs. We present 2 patients with atypical PSS, so-called "non-effort thrombosis," who were not involved in vigorous activities. They underwent thoracic outlet decompression through the infraclavicular approach without concomitant venoplasty. They were discharged without postoperative anticoagulant therapy. Venography and computed tomography after surgery revealed successful recanalization of the subclavian vein in each case. We highlight the characteristic pathophysiology of "non-effort thrombosis," an atypical PSS entity.

Keywords: Paget–Schroetter syndrome, deep vein thrombosis, effort thrombosis

Introduction

Paget–Schroetter syndrome (PSS) is an upper extremity thrombosis occurring in the axillary and subclavian veins (SCV).[1,2] PSS is a subtype of the thoracic outlet syndrome, in which surrounding neurovascular structures can be affected, accounting for variations in its presentations, including neurologic (95%), venous (4%), and arterial (1%).[3] PSS is also known as "effort thrombosis," because it is usually associated with repetitive and strenuous activities of the upper limbs. Although the diagnosis and management of PSS have improved significantly,[4] PSS without distinct causes, such as vigorous exercise, is rarely reported. Herein, we present 2 cases of atypical PSS, so-called "non-effort thrombosis," which were treated successfully by surgery using the infraclavicular approach.

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Case Report

Case 1

A 32-year-old woman, diagnosed with idiopathic organized pneumonia 4 months prior, was referred to our hospital. She complained of mild chest pain and repeated right arm pain with swelling. She was a healthy office worker without a smoking history and had no exercise habit. She was 164 cm tall and weighed 54 kg. Her general condition was good, except for the pain and swelling of the right arm, which worsened in the abduct position. A blood examination did not reveal protein C deficiency, protein S deficiency, and anti-phospholipid syndrome, and was negative for serum tumor markers and antinuclear antibodies. Computed tomography (CT) showed SCV thrombosis in the costoclavicular space (**Fig. 1a**) and peripheral pulmonary infarction as a result of recurrent pulmonary embolism. Venography revealed stasis of the right SCV and right axillary vein in the keyboard typing position (abduction to 30°); the stasis was absent in the resting position (**Fig. 1b**). Despite undergoing anticoagulant therapy with heparin and warfarin after thrombolysis with systemic infusion urokinase (480,000 U/day) for 1 week, the thrombosis recurred within a month. She decided to undergo first rib resection to improve the recurrent symptoms of PSS. Using the infraclavicular approach with a 6-cm incision, the first rib and SCV were exposed. The adhesion around the SCV was mild. The left costoclavicular ligament, anterior scalene muscle, and anterior

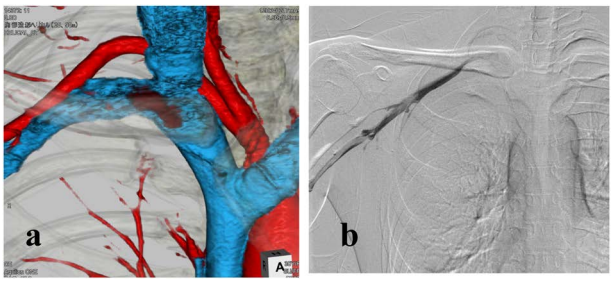

Fig. 1 Preoperative computed tomography and venography of patient 1.
(**a**) Computed tomography of patient 1 shows right subclavian vein thrombosis around the costoclavicular space. (**b**) The venography of patient 1 shows the compressed right subclavian vein and stasis of blood flow in the keyboard typing position (abduction to 30°).

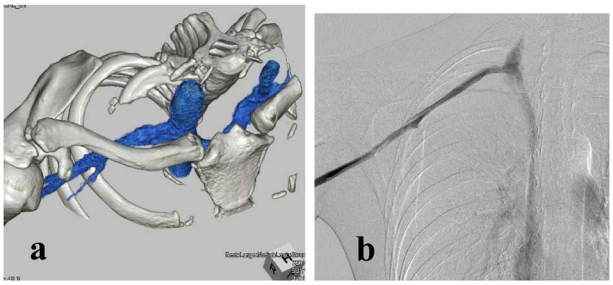

Fig. 2 Postoperative computed tomography and venography of patient 1 after the surgery.
(**a**) Postoperative computed tomography of patient 1 shows the right subclavian vein without stenosis and resected first rib. (**b**) Postoperative venography shows successful recanalization of the right subclavian vein in the keyboard typing position (abduction to 30°).

two-thirds of the first rib were resected. SCV reconstruction was not needed, because there was no venous wall degeneration causing stenosis. Her symptoms improved soon after the surgery. Venography and CT revealed successful recanalization of the right SCV at the keyboard typing position (**Figs. 2a** and **2b**). She was discharged at the fourth postoperative day without administration of anticoagulants. Her postoperative course was uneventful, and she had no recurrence of PSS 2 years postoperatively.

Case 2

A 36-year-old woman was referred to our hospital because of repeated swelling and pain in her left upper extremity during work. She was an office worker with no smoking history or family history of PSS. She also did not have an exercise habit. She was 161 cm tall and weighed 59 kg. As in case 1, no coagulopathy was noted and serum tumor markers and antinuclear antibodies were not elevated in the blood examination. CT and ultrasonography revealed a thrombosis in the left SCV. Venography demonstrated

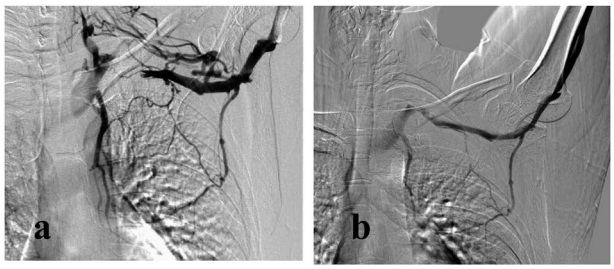

Fig. 3 Preoperative and postoperative venography of patient 2.
(**a**) The venography of patient 2 shows complete obstruction of the left subclavian vein with prominent collateral veins. (**b**) Postoperative venography of patient 2 shows successful recanalization of the left subclavian vein and loss of most collateral veins.

complete obstruction of the left SCV with the development of collateral veins (**Fig. 3a**), suggesting chronic SCV compression. She was diagnosed as having PSS and underwent anticoagulant therapy with systemic heparinization (15,000 U/day) for a day, followed by oral administration of dabigatran (300 mg/day) for 6 months. Although thrombolysis was achieved, her symptoms recurred within 2 months after finishing the anticoagulant therapy. She underwent first rib resection using an infraclavicular approach in the same manner as case 1. The patient did not need SCV reconstruction. Anticoagulant therapy was discontinued after the surgery. Venography revealed successful recanalization of the left SCV without major collaterals (**Fig. 3b**). She was discharged on the fourth postoperative day in good condition. Her postoperative course was uneventful, and she had no recurrence of PSS 1 year postoperatively.

Discussion

PSS is believed to be incited by repeated abnormal and strenuous interactions between the venous system and the structures within the thoracic outlet. PSS occurs frequently in young, active, and healthy patients. Most patients have a common history, such as frequent vigorous activity, repetitive overhead motion, and heavy lifting by the affected extremity.[1] However, the present patients did not have such histories. Our experiences highlight the presence of "non-effort thrombosis" without the typical manifestation of PSS. The main etiology of PSS is thought to be the chronic compression of the vein between the angle of the clavicle and the first rib with arm abduction. The narrowing costoclavicular space might be associated with PSS as well. The pathophysiology underlying PSS is chronic repetitive compression injury of the SCV, resulting in delayed sequelae of permanent SCV obliteration and disability.[1,5,6] Our patients worked in offices and spent a lot of time using the computer during the day. Venography

of case 1 revealed SCV occlusion in the keyboard typing position, suggesting that the SCV of non-effort cases has a narrower costoclavicular space and collapses easily in contrast to that in typical PSS. Of the 123 treated patients with PSS reported by Doyle and colleagues,[5] almost half of the patients did not have an obvious history of vigorous exercise, indicating that non-effort thrombosis may have been overlooked to date. A subgroup analysis or accumulation of experiences, focusing on this atypical PSS entity, is warranted.

The treatment of PSS consists of relieving symptoms due to obstruction, preventing complications from deep vein thrombosis, and preventing recurrence. This can be achieved through anticoagulation, thrombolysis, and/or surgical decompression. The initiation of systemic anticoagulation immediately after diagnosis is the first step of the treatment.[4] Anticoagulation alone for the treatment of PSS is generally not recommended. Hangge and colleagues reported in their review article that the aggressive approach involving thrombolysis and surgery was superior to anticoagulation alone in patient-reported outcomes, such as resolution of symptoms and return to work.[3] Anticoagulation poses another issue for young women with regard to the sexual cycle and/or the perinatal period. Medical personnel should take the above-mentioned information into consideration. Although there is no randomized control study on PSS, Vazques and colleagues reported a systematic review about primary deep arm vein thrombosis consisting of 25 studies and 1,271 patients.[6] In their article, the authors introduced a current guideline recommending anticoagulation therapy, although little has been discussed about postoperative anticoagulation therapy. Another review article reported that there was no consensus on the duration of anticoagulation for PSS.[3] In the present cases, stasis of SCV could not be seen even in the abduction position after the surgery, resulting in the anticoagulant-free non-recurrence of PSS. Our experiences highlight that surgical intervention with thoracic outlet decompression has the potential advantage of being an effective anticoagulant-free management for this atypical PSS entity. Moreover, postoperative venography is useful to determine whether the anticoagulant therapy should be continued.

The efficacy of emergency approaches to treat PSS has been reported.[7,8] However, implementing such approaches in all patients is difficult because of their heterogeneous physiological conditions. To prevent progressive venous scarring and stenosis, early surgery for PSS is recommended, although our patients took a few months from their original event caused by thrombosis to undergo surgery. Especially, case 2 seemed to have a long-term compression history of SCV because venography showed the development of collateral veins. Fortunately, their symptoms improved completely only by thoracic outlet decompression without concomitant venoplasty. This may suggest that the major factor of non-effort thrombosis is stasis, as described in Virchow's triad,[9] whereas that of effort thrombosis is endothelial injury. Given this pathophysiology, we consider that catheter hemolysis and anticoagulant therapy would sometimes be insufficient. Surgical intervention might be a better way to solve such situations. There are several surgical approaches to manage PSS. We chose the infraclavicular approach to allow surgeons to reach the SCV at the thoracic outlet directory for decompression and venoplasty, if needed, in 1 operative setting with minimal incision. Especially, the first rib and surrounding muscle or ligament resection, which is the most important step of this procedure, can be performed easily with this approach. Importantly, early surgery should be planned before the progression of SCV endothelial injury.

Conclusion

We experienced 2 cases of non-effort thrombosis treated by thoracic outlet decompression without venoplasty. These experiences suggested that a prompt approach was desirable for the treatment of this atypical PSS entity.

Disclosure Statement

All authors have no conflict of interest.

Author Contributions

Study conception: FS, RK, NK
Data collection: FS, RK, MC
Writing: FS
Supervision: RK, NK
Critical review and revision: all authors
Final approval of the article: all authors
Accountability for all aspects of the work: all authors

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