CASE REPORT

THE PATIENT
16-year-old boy

SIGNS & SYMPTOMS
– Pain, redness, and swelling of right upper arm
– No history of shoulder injury
– Positive Wright's and Adson's tests

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THE CASE
A 16-year-old boy presented to the emergency room (ER) with pain, redness, and swelling of his right upper arm that had been bothering him for 2 days. He was the quarterback of his high school football team, a sport that he'd been playing since he was 8 years old. He indicated that his football training—which involved repetitive throwing with his right arm—had intensified over the previous 2 months.

Prior to the ER visit, the patient was healthy and active with no significant medical history. He'd had no shoulder trauma and there was no family history of any coagulopathies, venous thrombosis, or pulmonary embolism. He denied chest pain, shortness of breath, palpitations, and fever, and said that he did not smoke cigarettes or drink alcohol.

On physical examination, his blood pressure was 118/70 mm Hg and his heart rate was 74 beats per minute. He had nonpitting edema and erythema of his right upper arm. His radial and brachial pulses were strong and equal in both arms. Assessment of neurologic and vascular integrity produced positive Wright's and Adson's tests, but a negative Halstead's test. (For more on these tests, see: https://youtu.be/gmtHStW7MB8 [Wright's test], https://youtu.be/pQw13a-kIDU [Adson's test], and https://youtu.be/gtGGCHR-y8I [Halstead's test].) The circumference of the patient's right upper arm was 2.5 cm greater than the left upper arm. The remainder of the physical exam was normal.

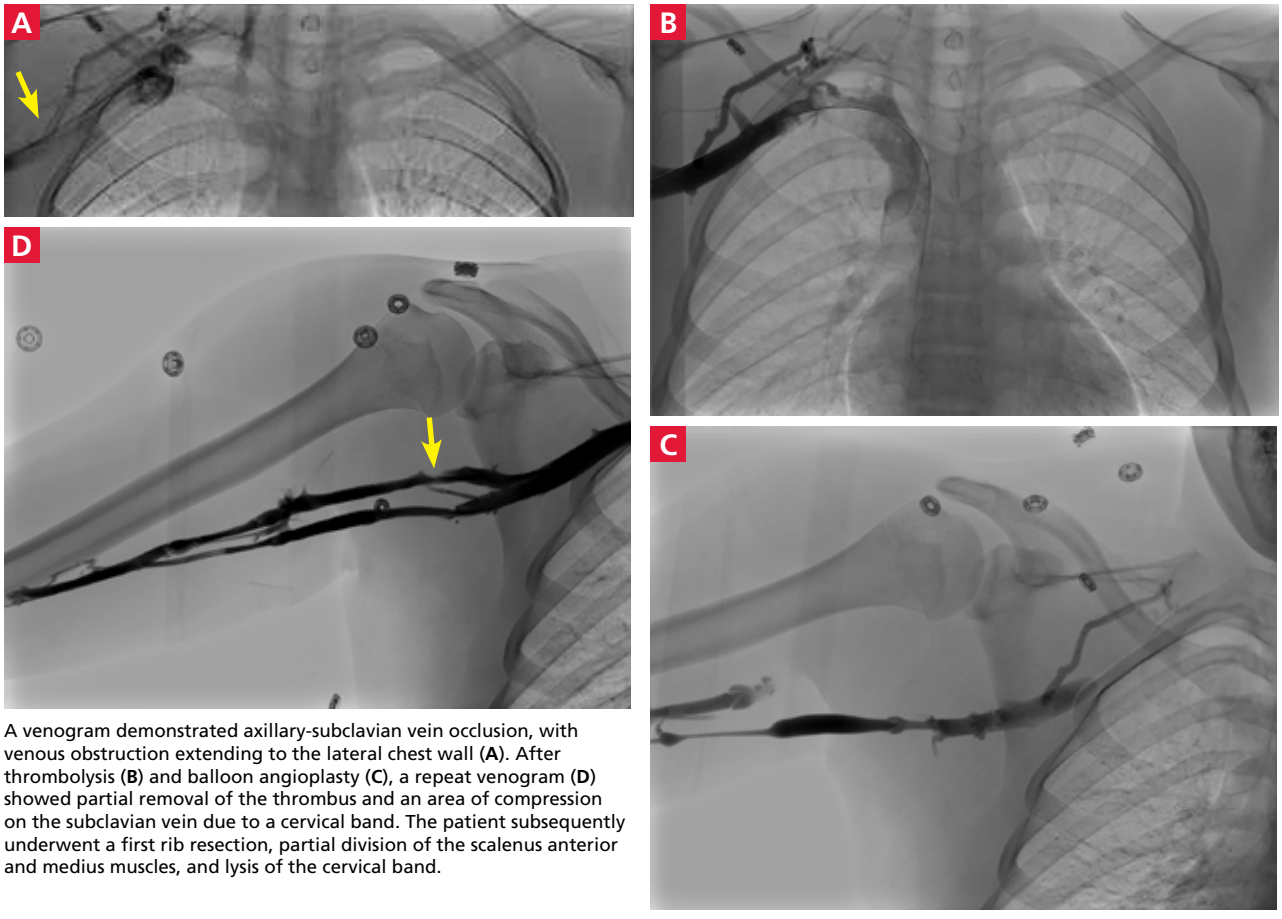
THE DIAGNOSIS
A duplex ultrasound of the right upper arm revealed an acute occlusive thrombus in the axillary vein. We started the patient on intravenous heparin. A venogram confirmed thrombosis of the axillary-subclavian vein (**FIGURE 1A**). Based on the patient's clinical presentation and the results of the venogram, we diagnosed Paget-Schroetter syndrome. The venogram was followed by thrombolysis with alteplase (**FIGURE 1B**) and a balloon angioplasty (**FIGURE 1C**). One week later, a repeat venogram demonstrated partial removal of the thrombus and an area of compression on the inferior aspect of the subclavian vein due to a cervical band (**FIGURE 1D**).

DISCUSSION
Paget-Schroetter syndrome (PSS), or effort thrombosis of the upper extremities, is defined as spontaneous thrombus in the axillary and subclavian veins that occurs as a consequence of strenuous upper-extremity activity. It is a rare condition with an incidence of one to 2 cases per 100,000 people per year, and represents 1% to 4% of all cases of deep vein thrombosis (DVT).[1]

Spontaneous thrombosis of the upper extremities typically presents in young, otherwise healthy individuals. It has been described in athletes who are involved in ball games,

FIGURE 1

Identification and management of Paget-Schroetter syndrome in a 16-year-old boy



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A venogram demonstrated axillary-subclavian vein occlusion, with venous obstruction extending to the lateral chest wall (**A**). After thrombolysis (**B**) and balloon angioplasty (**C**), a repeat venogram (**D**) showed partial removal of the thrombus and an area of compression on the subclavian vein due to a cervical band. The patient subsequently underwent a first rib resection, partial division of the scalenus anterior and medius muscles, and lysis of the cervical band.

games with rackets or clubs, aquatic sports, combatant sports, and in violin players.[2] The repetitive movements used in these activities can lead to compression of the axillary and subclavian veins by hypertrophied muscles. Repetitive trauma causes intimal damage and thrombogenesis.[3]

PSS is characterized by the abrupt, spontaneous swelling of the entire arm, cyanosis, and pain that occurs with use or overhead positioning. Enlarged subcutaneous veins are present in the upper arm, around the shoulder, or in the upper anterior chest wall (Urschel's sign). The classic presentation is acute onset of upper extremity pain and swelling in the dominant arm following a particularly strenuous activity.[4] A low-grade fever, superficial thrombophlebitis, or neurologic symptoms may coexist. Certain provocative maneuvers can help reproduce the symptoms (**TABLE 1**[5,6]). Complications of PSS include pulmonary embolism, postthrombotic syndrome (pain, heaviness, and swelling), and recurrent thrombosis.[7]

Contrast venography best shows the extent of thrombosis

Duplex ultrasound, with its high sensitivity and specificity, is the initial, noninvasive test of choice (**TABLE 2**[4,8-11]). However, duplex ultrasound has a false-negative rate of 30% because it is highly technician-dependent and can be complicated by acoustic shadows from the clavicle or sternum.[8]

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TABLE 1

Physical examination maneuvers for Paget-Schroetter syndrome[5,6]

Test	The maneuver *(Done in an effort to reproduce the symptoms/attenuate the brachial or radial pulse)*	Mechanism	Sensitivity (%)	Specificity (%)
Wright's test (hyperabduction maneuver)	The physician externally rotates and abducts the affected arm 180 degrees, with the elbow flexed 90 degrees, as the patient inhales deeply. The patient should turn his head to the contralateral side.	Compression of the neurovascular structures in the subcoracoid region.	90	29
Adson's test	The physician abducts the patient's affected arm 30 degrees while pulling it backward. At the same time, the patient takes a long breath while elevating his chin and turning it to the affected side.	Narrowing of the space between the scalenus anterior and medius, resulting in compression of the subclavian artery and the brachial plexus.	79	76
Halstead's test	The patient moves his shoulders inferiorly and medially, while sticking his chest out in a military posture.	Narrowing of the space between the first rib and the clavicle, thereby causing neurovascular compression.	84	47

The most direct and definitive means to confirm the diagnosis of PSS is catheter-directed contrast venography.[9] This method provides complete anatomic information regarding the site and extent of thrombosis, allows definitive evaluation of the collateral venous pathways, and is a necessary step toward the use of thrombolytic therapy. Contrast load, however, contraindicates the procedure in patients with renal failure and in those who are pregnant.

Contrast-enhanced computed tomography (CT) and magnetic resonance angiography (MRA) are also highly sensitive for detecting focal stenosis at the level of the first rib, the presence or absence of enlarged collateral veins, and the chronicity of any thrombus present. However, the usefulness of CT and magnetic resonance venography in initial screening is unclear, due to a lack of randomized controlled trials.

Treatment involves anticoagulants, thrombolytics, and possibly surgery

Prompt use of anticoagulation is indicated in PSS. Initial anticoagulation with low molecular weight unfractionated heparin or a direct thrombin inhibitor followed by warfarin for a minimum of 3 to 6 months is recommended.[12]

Patients treated with anticoagulation alone have a higher incidence of long-term residual symptoms, disability, and recurrent thrombosis.[7] As a result, a more aggressive approach with the use of thrombolytic therapy is indicated, especially in young, active patients, to minimize long-term consequences. Alteplase or reteplase are used for this purpose. Thrombolysis is less likely to be beneficial if the thrombus is more than 2 weeks old or if there are inflammatory changes in the vein. The use of catheter-directed thrombolysis minimizes the risk of systemic adverse effects and achieves higher clot resolution rates.[13]

Because PSS is caused by compression of the vein, rather than a disorder of blood clotting, there is still a 50% to 70% risk of recurrent thrombosis despite thrombolysis and anticoagulation.[14] Therefore, the most definitive management approach remains surgical treatment. Patients with recent thrombosis who are within the first several weeks of undergoing successful thrombolytic therapy are excellent candidates for surgery. Operative treatment for PSS includes first rib resection, scalene muscle removal, or subclavius mus-

TABLE 2

Radiologic studies for Paget-Schroetter syndrome[4,8-11]

Test	Advantages	Disadvantages
Duplex ultrasound	Sensitive and specific for peripheral vein thrombosis (observational studies) Noninvasive	Technician dependent Low specificity for anatomic landmarks Cannot use for exclusion due to false-negative rate of 30%
Catheter-directed contrast venography	Complete anatomic information regarding the site and extent of thrombosis and evaluation of the collateral venous pathways Required for catheter-directed thrombolytic therapy	Invasive Contraindicated in renal failure and pregnancy, due to the contrast load Technically difficult to introduce a catheter into an edematous arm
Contrast-enhanced CT scan or MRA	Highly sensitive for detecting anatomic problems Noninvasive	Insufficient data to support role as initial screening test due to lack of RCTs

CT, computed tomography; MRA, magnetic resonance angiography; RCTs, randomized controlled trials.

cle removal, along with removal of constricting scar tissue from around the vein.[7]

THE TAKEAWAY

PSS is characterized by upper-extremity DVT resulting from repetitive trauma to the subclavian-axillary vein. Early diagnosis of PSS with contrast venography and prompt use of anticoagulation can effectively restore venous patency, reduce the risk of rethrombosis, and return the patient to normal function. Primary care physicians should be aware of this condition, because delayed recognition in a high-functioning person can be potentially disabling.

Our patient had a first rib resection, partial division of the scalenus anterior and medius muscles, and lysis of the cervical band. Follow-up venography confirmed resolution of thrombosis without any complications. The patient was continued on anticoagulation with warfarin for 3 months. JFP

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