Diagnostic Imaging

Paget-Schroetter syndrome with bilateral pulmonary emboli

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ABSTRACT

Paget-Schroetter syndrome, also known as effort thrombosis, is a relatively rare disease process characterized by primary thrombosis to the subclavian and axillary veins. It usually presents in younger individuals, commonly affecting the dominant side upper extremity, and the diagnosis relies on a combination of imaging, laboratory tests, and clinical presentation. Upper extremity deep vein thrombosis can also lead to pulmonary emboli, as in this case of a 20-year-old female discovered to have right sided Paget-Schroetter syndrome.

Case report

A 20-year-old female with history of right shoulder bursitis presented to the emergency department experiencing pain in her right arm for 5 days, more severe to the shoulder and axilla, and swelling for 4 days. Blue discoloration of her right arm was elicited when she raised her limb above her head or lifted heavy objects. She did not describe chest pain or shortness of breath. There was no recent trauma or hospitalizations. Her only medications were lisdexamfetamine dimesylate for attention deficit disorder and an oral birth control. On physical examination, her right arm was swollen with a slight bluish discoloration, and her range of motion was limited by pain. Distal extremity pulses were palpable bilaterally.

Initial radiographs of the patient's chest and right shoulder were unrevealing. A right upper extremity venous duplex ultrasound examination found an occlusive thrombus and absent Doppler flow in the right subclavian and axillary veins (Fig. 1). Computed tomography angiography of the chest revealed bilateral filling defects of the pulmonary arteries consistent with thromboembolic disease at the lower lobes, such as at the bifurcation of the proximal right lower lobe segmental arteries (Fig. 2) and in the posterior basal branches of the left lower lobe. Evidence for a right subclavian venous clot was also noted on computed tomography (Fig. 3). Coagulation studies were all within normal limits. Although her oral contraceptive use may have been a contributing factor, the extent of thrombus hinted at another underlying etiology.

The patient was started on intravenous unfractionated heparin. Subsequent venogram confirmed an interruption of contrast flow to the right axillary and subclavian veins consistent with an occlusive thrombus (Fig. 4). A wire was used to traverse the occlusion and inject the superior vena cava, which was widely patent. These findings suggested Paget-Schroetter syndrome. An infusion length EKOS lysis catheter was then placed and thrombolysis with tissue plasminogen activator was initiated. A repeat venogram the next day showed...
significant resolution of the thrombus with narrowing of the subclavian vein at the thoracic outlet (Fig. 5). The sheath was removed and patient was transitioned to oral anticoagulant rivaroxaban on discharge. As part of the patient’s management plan, about 2 months later she underwent a transaxillary resection of the right first rib with right subclavian vein balloon angioplasty and deployment of a 14-mm self-expanding stent. Subsequent venogram demonstrated patent right axillary and subclavian veins. Surgical exploration found impingement of the vein was due to medial attachment of the anterior scalene muscle and a thick fibrous costoclavicular ligament, confirming the diagnosis. Postoperative chest radiograph demonstrated the rib resection and stent (Fig. 6). Follow-up Doppler ultrasound 1 month after demonstrated a chronic but nonocclusive thrombus in the right subclavian and axillary veins. The stent in the subclavian vein was patent (Fig. 7), and the patient endorsed resolution of her previous symptoms.

Discussion

Deep venous thrombosis of the upper extremity accounts for only 10% of all documented cases of deep vein thromboses (DVTs) with an annual incidence in the general population from 0.4 to 1 cases per 10,000 [1]. Paget-Schroetter syndrome, also called effort thrombosis, is a relatively rare disease process and refers to primary thrombus formation in the subclavian and
axillary veins, separate from the much more common catheter-related thrombosis. It usually presents in younger individuals with a predominance for males and commonly affects the dominate side upper extremity [2]. The pathophysiology is believed to be a product of overuse and strenuous activity combined with anatomic abnormalities. This diagnosis is the venous subtype of thoracic outlet syndrome, which broadly also includes compression of the brachial plexus or axillary artery. The anatomic variants usually involve abnormal costoclavicular ligament enlargement and hypertrophy of the anterior scalene muscles leading to a narrowing of the costoclavicular space, creating obstruction within the pathway of the subclavian vein [2]. The immobilization of the vein due to extrinsic obstruction leads to a higher chance of injury from normal strenuous physical activity. Because these compressions of the subclavian vein are usually chronic in nature, repeat injury occurs and can eventually lead to fibrotic development within the vessel. This results in additional stenosis and obstruction of venous blood flow [2].

Fig. 4 – Right upper extremity venogram demonstrates obstruction of contrast to the axillary and subclavian veins with collateralization consistent with occlusive thrombus.

Fig. 5 – Follow-up right upper extremity venogram now demonstrates patent contrast flow to the axillary and subclavian veins. Narrowing of the subclavian vein is present at site of previous thrombus.

Fig. 6 – Portable AP chest radiograph shows postsurgical changes from a right first rib resection and stent in the proximal right subclavian vein. A right pneumothorax is also present (arrow). AP, anteroposterior.

Fig. 7 – Follow-up venous Doppler ultrasound of the right upper extremity shows patency of the right subclavian vein stent.
Evaluation for Paget-Schroetter syndrome relies on a combination of imaging, blood tests, and clinical symptoms to make the diagnosis. Common nondescript symptoms include pain, swelling, discoloration of the affected extremity, as well as general limb discomfort. However, clinical presentation alone has a poor predictive power yielding specificity less than 50% \[2\]. Initial plasma D-dimer testing has a relatively high sensitivity but lacks specificity as many inflammatory processes can also lead to elevated D-dimer levels \[3\].

The most appropriate first test is an ultrasound because it is noninvasive and inexpensive. Doppler ultrasound also has a relatively strong diagnostic power with sensitivity for DVTs range from 78% to 100% and specificity range from 82% to 100% \[4\]. X-ray venography remains the most accurate imaging test available and is the gold standard; however, because of the invasive technique, it is often reserved for situations of high clinical suspicion, but when other tests were unable to confirm the presence of an upper extremity DVT \[5\]. Contrast-enhanced venography is also widely utilized before and after lysis therapy or surgery to assess the status of a known thrombus. An upper extremity DVT on an x-ray venogram is illustrated by a filling defect, an abrupt change of the flow of contrast and enlargement of collaterals around the suspected obstruction \[3\]. Pulmonary embolism is a known complication of DVT development and occurs less frequently with DVTs in the upper extremity (6%) compared with the lower extremity (15%-32%) \[1\].

The management of patients with Paget-Schroetter syndrome can involve 4 components: anticoagulation therapy, catheter directed thrombolysis, mechanical catheter interventions, and surgery. The initial regimen should consist of low molecular weight heparin for at least the first 5 days from presentation with eventual transition to an oral anticoagulant for an additional 3-6 months \[1\]. Catheter-directed thrombolysis utilizing tissue plasminogen activator directly instills thrombolytic agents to the site of the thrombus. Surgical intervention is generally indicated when the patient is symptomatic from DVTs in the affected extremity, a venogram is positive for significant compression, and following thrombolytic therapy \[6\]. Decompressive surgical resection of the first rib after thrombolysis has an expected favorable clinical outcome in 80%-90% of patients \[7\]. Mechanical catheter interventions include catheter-directed thrombectomy, balloon angioplasty, and stenting. Evidence has shown that angioplasty and stenting without decompressive surgery have high failure rates \[6\] as the tight costoclavicular junction persists.

### Conclusion

Paget-Schroetter syndrome should be considered for an axillary and subclavian DVT if there are no other underlying reasons like prior central catheter or pacemaker insertion. It is attributed to repetitive motion and anatomic compression of the subclavian vein at the thoracic outlet. Oral contraceptive use such as in this patient may have been a contributing factor too. There is also a small risk for concurrent pulmonary emboli. Because venous thrombi will recur even after catheter-directed thrombolysis, the long-term treatment plan often involves surgical resection of the first rib and the crowding structures around the subclavian vein.

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