Paget-Schroetter syndrome in pregnancy: A case report and discussion of management options

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ABSTRACT

Paget-Schroetter syndrome is a form of primary venous thoracic outlet syndrome whereby thrombosis of the subclavian vein is instigated by repeated and vigorous overhead activity of the arm. We describe a 35-year-old pregnant white woman at 8 weeks' gestation, a competitive swimmer by profession, who was diagnosed with a left upper extremity thrombus. First rib resection through an infraclavicular approach was performed 1 week after percutaneous pharmacomechanical thrombectomy. Repeated venography demonstrated residual thrombus requiring percutaneous pharmacomechanical thrombectomy with balloon venoplasty. We present a challenging case with focus on the unique diagnostic evaluation and management of pregnant patients with this condition. (J Vasc Surg Cases and Innovative Techniques 2020;6:59-62.)

Keywords: Paget-Schroetter syndrome; DVT; Pregnancy; Vascular interventions; Surgery

Thoracic outlet syndrome (TOS) refers to a set of symptoms caused by the compression of the subclavian vein or artery or the brachial plexus. Paget-Schroetter syndrome (PSS) refers to spontaneous venous thrombosis. The most common cause is venous presentation of TOS, accounting for 4% of all cases of TOS; it is thought to be primarily due to repeated, vigorous overhead activity of the arm causing venous obstruction.[1,2] In pregnant patients, this risk of deep venous thrombosis (DVT) is even higher; women who are pregnant or post partum have a fourfold to fivefold increased risk of thromboembolism compared with nonpregnant women.[3] Initially, most centers perform catheter-directed thrombolysis and an interval first rib resection a few days after thrombolysis if extrinsic compression of the subclavian vein at the thoracic outlet is demonstrated.[4,5] Whereas data exist on the efficacy of thrombolysis and interval surgical decompression, consensus regarding the comprehensive management of patients with PSS is lacking because of the paucity of prospective, randomized studies delineating definitive outcomes, much less in those who are pregnant. Thus, special consideration should be given to pregnant patients with PSS for whom medical and surgical intervention is warranted. We report successful treatment of this rare condition in a pregnant patient with PSS presentation of venous TOS, who has given informed consent to present the case.

CASE REPORT

A 35-year-old G1P0 white woman at 8 weeks by last menstrual period, a competitive swimmer by profession with no significant medical history, presented to an outside facility with a 2-day history of acute-onset left upper extremity paresthesia, discoloration, and swelling after swimming for 90 minutes. Initial Doppler ultrasound performed at an outside imaging center revealed venous thrombus extending from the left brachial to the axillosubclavian vein with a patent left internal jugular vein and a viable pregnancy. Physical examination at presentation to our center was significant for grossly swollen left upper extremity, bluish discoloration of distal left phalanges and forearm, and engorged superficial upper arm veins with palpable radial and ulnar pulses bilaterally. Therapeutic anticoagulation with heparin drip was initiated at presentation before changing to enoxaparin on hospital admission day 1. Before planning operative intervention, the obstetrics service was consulted for recommendations concerning safe anticoagulation for venous thromboembolism during pregnancy and re-evaluation of the fetus.

On hospital admission day 2, the patient underwent left upper extremity venography with percutaneous pharmacomechanical thrombectomy (PMT) using AngioJet (Boston Scientific, Marlborough, Mass). Care was taken to minimize radiation exposure to the patient and fetus by placing a lead apron on the operating table and over the patient's abdomen. After the left brachial vein was identified and an 8F sheath was inserted for access, the occluded axillosubclavian vein was traversed using a 0.035-inch stiff Glidewire (Terumo Interventional Systems, Somerset, NJ) and an angled Kumpe catheter. Subsequently, venography demonstrated subacute thrombus extending from the axillary vein to the distal left subclavian vein. This was confirmed with intravascular ultrasound, which also demonstrated

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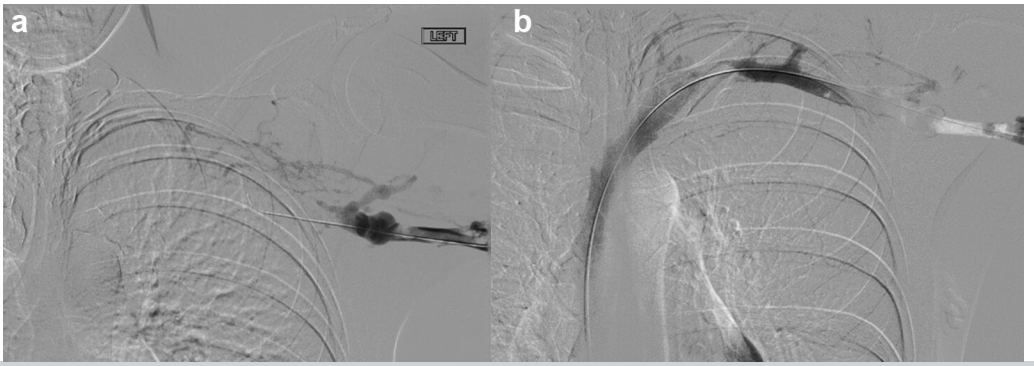

Fig 1. a, Digital subtraction venogram of the left upper extremity revealing deep venous thrombus originating at the axillary vein. **b,** Post-AngioJet digital subtraction venogram of the left upper extremity revealing improved flow of contrast material through a compressed thoracic outlet to the superior vena cava.

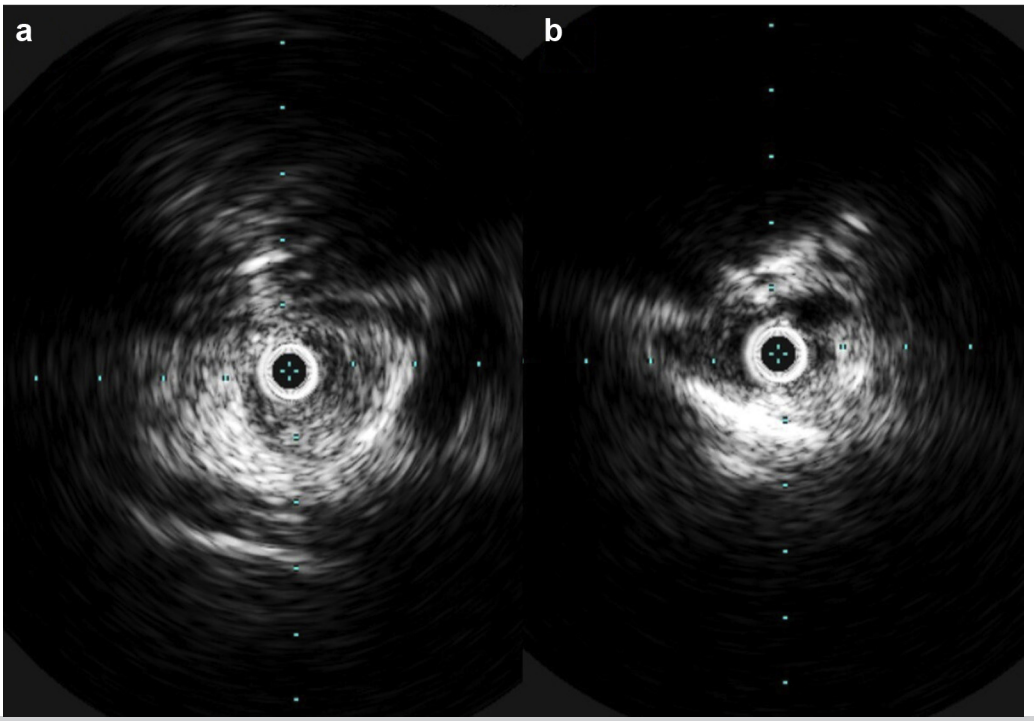

Fig 2. Intravascular ultrasound images of the subclavian vein before **(a)** and after **(b)** AngioJet demonstrating improvement in the subacute thrombus burden after pharmacomechanical thrombectomy (PMT).

significant venous wall scarring (Fig 1, *a*). An AngioJet catheter was then advanced to perform PMT and to pulse spray tissue plasminogen activator (tPa). Completion venography revealed 50% clot resolution and brisk flow of contrast material through the axillosubclavian vein into the superior vena cava with residual compression of the left subclavian vein within the thoracic outlet where it traversed the first rib, confirming the diagnosis (Figs 1, *a*, and 2). There were no postoperative complications. Surgical intervention to relieve compression of the thoracic outlet through first rib resection was advised to prevent further recurrence of DVT and venous scarring complications.

The patient was then discharged from our service to present 5 days later for an elective rib resection. A bedside fetal ultrasound examination was performed before and after surgery, which confirmed viable pregnancy. Through a left infraclavicular approach, the subclavian vein was identified and dissected free and mobilized. The left subclavian vein was visibly compressed between a hypertrophic subclavius muscle and the first rib and clavicle. After the subclavius muscle was excised at its junction with the sternoclavicular joint, the subclavian vein was dissected free, and the first rib was then mobilized by dividing the intercostal muscle attachments and anterior scalene muscle (Fig 3). A rib cutter was used to divide the rib both inferiorly and superiorly. During the same operation, venography was performed, which demonstrated persistent filling of collaterals with minimal flow through the subclavian vein. Repeated percutaneous PMT with AngioJet was initiated, followed by balloon venoplasty with a 12-mm Conquest balloon (Bard, Murray Hill,

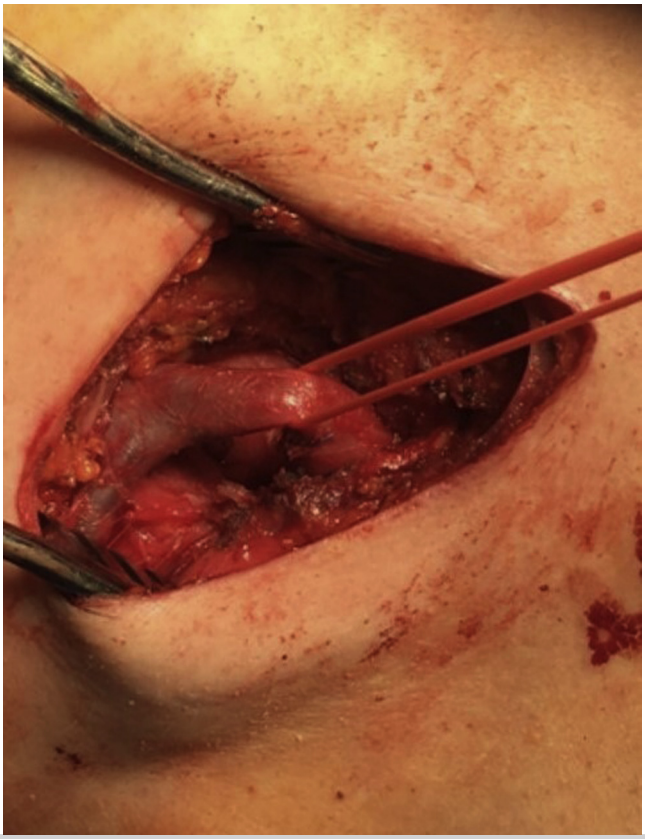

Fig 3. Operative photograph of the thoracic outlet demonstrating mobilization of the subclavian vein after removal of the first rib.

NJ) with satisfactory result. The patient was prescribed therapeutic enoxaparin 1 mg/kg twice daily for the remainder of pregnancy. On a follow-up appointment 6 months postoperatively, physical examination findings were improved to baseline, and the patient had no other complaints. A left upper extremity venous duplex ultrasound evaluation revealed widely patent left subclavian and axillary veins with minimal thrombus.

DISCUSSION

Our literature review returned several reports of PSS in women of childbearing age, of whom most had a history of repeated, vigorous ipsilateral upper extremity use.[6-8] However, we could not identify any description of the clinical presentation and management of PSS in the pregnant patient.[9,10]

Although multiple surgical and nonsurgical approaches have been used for the treatment of PSS, most patients are now treated with a combined approach of PMT and surgical decompression with long-term anticoagulation of at least 3 to 6 months. We opted for a staged combination therapy for our patient using initial anticoagulation, followed by catheter-directed thrombolysis, and finally first rib resection to definitively treat the patient and to prevent thrombus progression or recurrence during her pregnancy.

Pregnancy induces hypercoagulability. Several mechanisms have been proposed for this state, including increased coagulation factors and venous stasis due to a systemic decrease in venous tone.[11-13] Hence, in our patient, there were two clearly identifiable risk factors (pregnancy and chronic venous injury) that contributed to the development of PSS with acute progression. As there is a lack of guidelines on treating PSS in the pregnant patient, we chose to offer a combination of already established treatment options, paying special attention to fetal monitoring and maternal safety interventions. We based our anticoagulation choice of therapeutic enoxaparin on established guidelines with no risk of teratogenicity and low risk of heparin-induced thrombocytopenia, as opposed to warfarin and direct oral anticoagulants, which have known teratogenic and embryo-fetal concerns, respectively.[14-17] The other consideration is the administration of local tPA while performing PMT in the pregnant patient; tPA is considered to be a class C drug and can be used during pregnancy without significant increase in maternal or fetal complications.[18] In this particular case, local rather than systemic administration of tPA confers an even smaller risk. Whereas some authors suggest that a 3-month interval between recanalization and surgical decompression confers adequate symptom relief, rethrombosis can occur in up to 10% of patients during this interval.[19] To mitigate this complication, an early or prompt first rib resection has been seen to have the same complication rates as later decompression with excellent outcomes. Given her clot burden and the early stage of her pregnancy, it was deemed safe to offer her surgical decompression 5 days after discharge instead of waiting and risking recurrence of the DVT and possibly occurrence of pulmonary embolism during the later stages of pregnancy. With regard to duration of anticoagulation status after rib resection, we elected to continue this for the remainder of her pregnancy, given the known relative increased risk of hypercoagulability associated with pregnancy. On a follow-up appointment 6 months postoperatively, at which point she was immediately post partum, duplex ultrasound examination revealed widely patent subclavian vein, and thus anticoagulation was discontinued.

CONCLUSIONS

Treatment of PSS in the pregnant patient presents several unique considerations. First, the choice of anticoagulation must be safe for the fetus. In addition, the gestational age of the fetus is a strong consideration in planning of definitive surgery. The potential risks and benefits of long-term anticoagulation, especially in the peripartum period, should be evaluated alongside the risk for development of worsening thrombosis, especially in the last half of pregnancy. Whereas our report focuses on the successful treatment of PSS in a pregnant patient, the management of anticoagulation and thrombolysis in pregnancy should be further investigated for this high-risk population.

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