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Review Article

May–Thurner and Paget–Schroetter Syndromes: A Review

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May–Thurner and Paget–Schroetter syndromes are rare conditions encountered by vascular surgeons. An updated knowledge about these conditions is crucial for the effective management of patients with these syndromes who are mostly young.

May–Thurner syndrome (MTS) is caused by the compression of the left common iliac vein by the right common iliac artery, and it is a risk factor for left leg deep venous thrombosis (DVT). Imaging (conventional venogram, computed tomography venography (CTV), magnetic resonance imaging (MRV)) can reveal the stenotic venous segment where the artery crosses. Stenting in symptomatic patients yields good results with minimal recurrence.

Paget–Schroetter syndrome (PSS) is an idiopathic axillary-subclavian vein thrombosis mostly affecting the young population, particularly those who have repeated overhead arm activities. Narrower costoclavicular space along with other anatomical and coagulation factors can possibly lead to this condition. Patients can present with acute DVT, post-thrombotic syndrome or subclinical syndromes. Venogram, CTV or MRV with provocative maneuvres can confirm the dynamic obstruction. Thrombolysis followed by early bony decompression is advocated in patients presenting with acute arm DVT, as this approach is associated with limited residual disability and recurrence.

This review discusses the current concepts and treatment options of both conditions.

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May–Thurner Syndrome

May–Thurner syndrome (MTS) is a condition in which the left common iliac vein is compressed by the right common iliac artery against the vertebra. It increases the risk of deep venous thrombosis (DVT) in the left leg. It was first described in 1957 by May and Thurner in an autopsy report of 430 cadavers, of which 22% had this condition.[1] This variant has been shown to be present in over 20% of the population; however, it is rarely considered in the differential diagnosis of DVT.

Aetiology

Compression causes 'intimal hyperplasia' which creates the potential site for venous stasis and subsequent thrombosis. The overlying artery may partially obstruct the left common iliac vein and damage the intima by its chronic pulsations. This condition has been estimated to occur in 2%–5% of the patients undergoing evaluations of the lower extremity venous disorders, and it remains unknown why the normal anatomical relationship between the left common iliac vein and right common iliac artery is disturbed and begins to interfere with venous flow.[2]

Clinical Presentations

The classic presentation of MTS is a young female in the second or third decade of life presenting with acute left lower extremity swelling involving the entire limb. Kim et al. described the three stages of iliac vein compression: stage I, asymptomatic iliac vein compression without any narrowing; stage II, when there is a development of venous spur without thrombosis; and Stage III, when there is left iliac vein DVT.[3]

Thurner et al. have advocated the use of pressure differential to support the diagnosis of hemodynamically significant obstruction. They have suggested that a differential pressure between two iliac veins of 2 mmHg at rest or 3 mmHg with exercise is significant and that an exaggerated pressure response to exercise is a marker of a

significant obstruction.

Investigations

Compression ultrasound is chosen for diagnosing DVT, but it has limitations in visualising MTS that occurs high in the pelvis.[4] MTS diagnosis requires the demonstration of the stenotic or occlusive venous segment on imaging, such as contrast venography, magnetic resonance imaging or intravenous ultrasound[5,6] (**Fig. 1**). The 'gold standard' for MTS diagnosis is venography, which can be both diagnostic and therapeutic. Intravenous ultrasound (IVUS) is also highly sensitive and specific in delineating morphology of the 'spur' and degree of the stenosis.

Treatment

Anticoagulation is not adequate to prevent long-term sequelae in patients with MTS[7]; a more invasive therapeutic approach is indicated.[8] Surgical procedures, such as repositioning of overriding vessel and veno-venous bypass, are of historical interest. After the first case report of successful venous stenting to relieve iliac obstruction,[9] several subsequent studies demonstrated the efficiency of thrombectomy and endovascular stenting in MTS, with 2-year iliac vein patency rates from 95% to 100%.[10] Following stent placement, systemic anticoagulation is recommended for at least 6 months.[11] Treatment is not recommended for asymptomatic patients. For moderate to severe symptomatic patients with MTS in the absence of DVT, venoplasty and stenting of the affected segment is recommended[11] (**Fig. 2**). For suspected MTS with DVT, catheter-directed thrombolysis or pharmacomechanical thrombolysis along with anticoagulation, followed by stenting, is recommended.[12] Self-expanding stents cover a long distance, have adequate durability and are preferred in this situation.[13] Balloon expandable stents may be used if needed (insufficient response to pre-dilatation and to contour self-expandable stent).

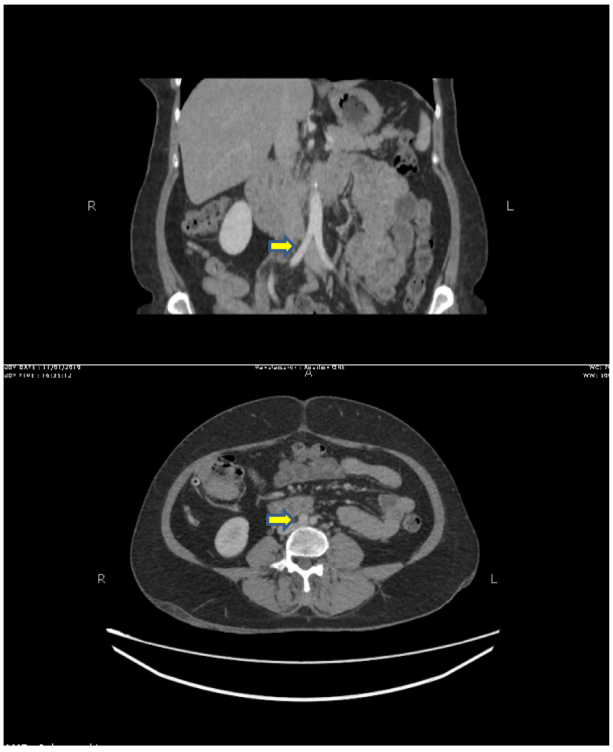

Fig. 1 A 48-year-old male presented with left leg venous ulcer. Computed tomography venography (CTV) reveals compression of the left common iliac vein by the right common iliac artery (May–Thurner syndrome).

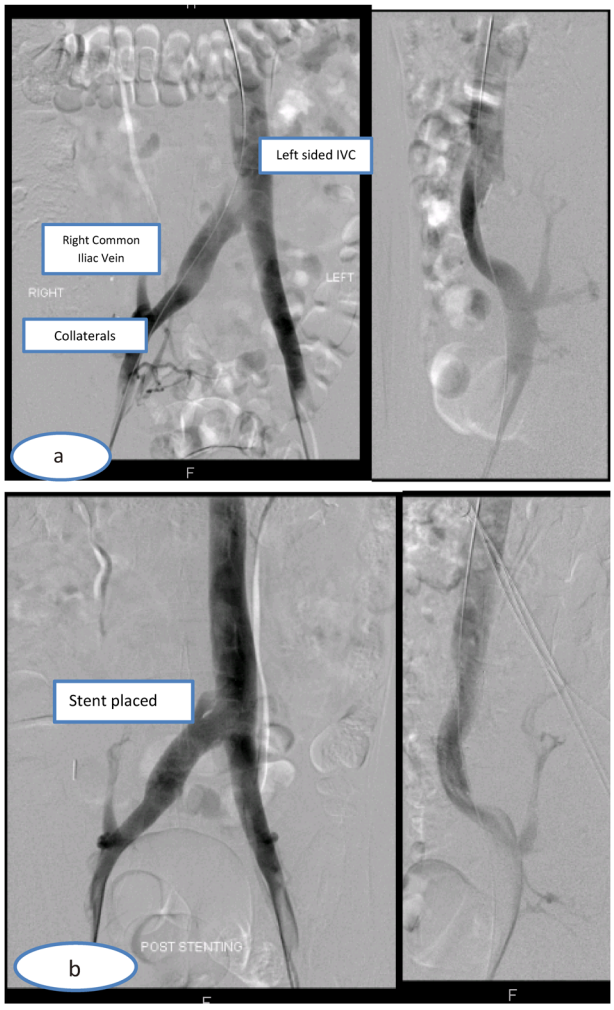

Fig. 2 A 28-year-old female with acute right leg swelling. Venogram revealed narrowing of the right common iliac vein with multiple collaterals and left-sided inferior vena cava (a variant of May–Thurner syndrome). The patient was in supine position (**a**). A self-expanding stent (14×50 mm) placed across the stenotic segment (**b**). The final venogram revealed marked improvement in the flow with reduced collaterals (**b**).

Paget–Schroetter Syndrome

Paget–Schroetter syndrome (PSS) is an idiopathic thrombosis of axillary and subclavian veins. It is one of the main causes of upper-arm DVT in young individuals without any predisposing factors.[14] It is also known as 'effort thrombosis' affecting individuals involved in physical activities,[15] such as athletes, wrestlers and those playing ballers. The costoclavicular space is a rigid and narrow space from where the subclavian-axillary artery passes. Repeated arm activities can damage the adventitia and induce microtrauma to the intimal wall of the vein. Perivenous inflammation and later fibrosis lead to vein narrowing and shortening. The shortened vein is even more at risk of tearing with repetitive movements. Anatomical abnormalities involving the thoracic inlet (cervical rib, congenital bands, hypertrophy of the scalene muscle and abnormal insertion of costoclavicular ligaments) and even the underlying subclavius muscle can compress it.[16] Although considered idiopathic, hypercoagulable conditions have been reported in 67% of the patients[17] (**Fig. 3**).

Clinical Presentations

The most common clinical presentations are arm swelling and arm discomfort.[18] Others can be cyanosis and dilated visible veins across the shoulder and upper arm (Urschel's sign). Patients presenting with intermittent venous obstruction only have arm swelling or normal physical examination, and the occlusion is evident on the provocative tests on venogram or computed tomography venography (CTV)/magnetic resonance imaging (MRV). Subclinical syndrome is caused by repeated partial thrombosis; patients may or may not be symptomatic and may also present with complications, such as pulmonary embolism and post-thrombotic syndrome.

Diagnosis

Although ultrasound is the investigation of choice for the diagnosis of DVT, it cannot directly diagnose the compression of the axillary-subclavian vein at the costoclavicular junction. Central venography has been the gold standard for the diagnosis of PSS (**Fig. 4**). MRV and CTV are alternatives with high sensitivity (100%) and specificity (97%). A provocative test (hyperabduction) helps confirm the dynamic obstruction.[19]

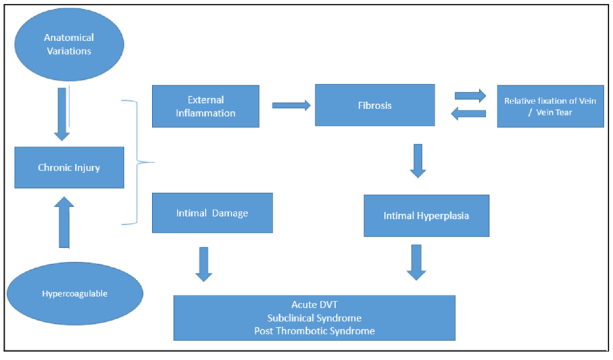

Fig. 3 Pathophysiology of Paget–Schroetter syndrome.

Management

These are the questions that come to mind while managing these patients:

Is conservative treatment alone sufficient in dealing with this condition?

What is the best method for treating the thrombus?

What are the most appropriate time and method to correct bony compression?

How to deal intrinsic venous defects?

1. Is conservative treatment alone sufficient in dealing with this condition?

Conservative treatment (anticoagulation and limb elevations) alone is not effective to provide complete symptomatic relief in these patients. Residual upper-arm obstruction and persistent symptoms are present in 29% to 68% of the patients.[20,21]

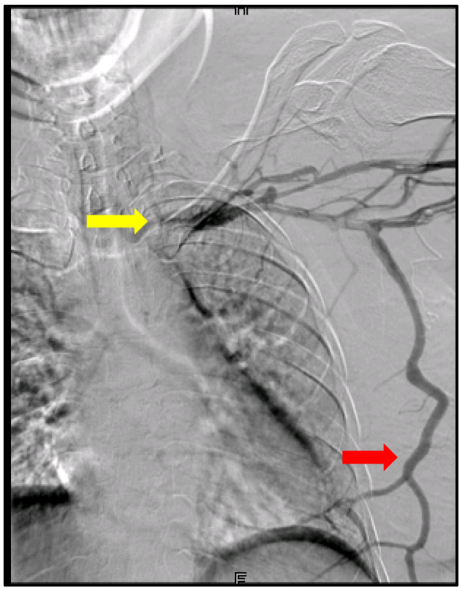

Fig. 4 A 35-year-old female presented with swelling in the left shoulder, supraclavicular fossa and breast for the last 6 months. She never had central venous cannulation. Upon examination, left-arm swelling with the appearance of prominent veins was observed. Venogram revealed features of chronic deep venous thrombosis with occlusion at the costoclavicular level (yellow arrow) and large collaterals (red arrow).

2. What is the best method for treating the thrombus?

Catheter-directed thrombolysis not only dissolves the clot but also preserves the intimal lining of the veins. It has become the standard first step for all patients with acute effort thrombosis unless significant contraindications are present. Successful recanalisation had been reported in 62%–84% of the treated cases.[22] This rate is even higher for patients with fresh clot approaching 100%, if initiated within a few days of symptom onset. It also reduces the risk of pulmonary embolism. The young active individuals are the ones most affected, and a minor degree of disability significantly affects their day-to-day activities and result in their poor quality of life. Thrombolysis is more effective if clot burden is less and it is relatively fresh (< 14 days).

3. Is bony decompression necessary?

Thoracic outlet decompression has been advocated in patients with persistent or recurrent symptoms following catheter-directed thrombolysis. Lee et al. used this strategy and reported that less than 25% of patients required surgery after a mean follow-up of 13 months.[23] Others recommend routine and early decompression in all patients.[24]

4. What is the most appropriate time and method for correcting bony compression?

If surgical decompression is not performed, rethrombosis is reported to occur within 30 days in as many as one third of patients. Venoplasty and stenting, although tempting, is not a reasonable option in this situation. Stenting in the costoclavicular junction without decompression is associated with stent fracture and deformation.[25] Both transaxillary and supraclavicular approaches can be used for decompression. Transaxillary approach is cosmetically appealing but technically challenging, and most vascular surgeons are not familiar with this approach. It provides adequate exposure to the anterior portion of the first rib. The potential complications are damage to the lateral thoracic nerve, hemothorax and pneumothorax and potential excision of the 'second rib' by an inexperienced operator. Transaxillary first rib resection provides 'good to excellent' long-term results in 85% to 95% of the patients. Supraclavicular/paraclavicular approach also provides good exposure for decompression 'under vision,' but it is cosmetically inferior to the transaxillary approach. Whatever approach is used, it is absolutely critical to address the anterior first rib and/or medial clavicle, and the vein must be freed from surrounding fibrotic tissues and be well mobilised.

Decompression can be performed 'early' during hospital stay or as 'staged,' months after thrombolysis. The problem with 'staged reconstruction' is that rethrombosis can occur in more than 10% of the patients within this interval.[26] Urschel and colleagues were proponents of early decompression after thrombolysis. They reported good or excellent results in 95% of 199 extremities treated within 6 weeks of symptom onset with no recurrences.[16] Molina et al. demonstrated the safety of immediate supraclavicular first rib resection after thrombolysis in 97 patients with only one bleeding complication.[24]

5. How to Deal with Intrinsic Venous Defects? Percutaneous or Surgical Venoplasty

About one third of the patients have intrinsic vein defects after thrombolysis.

The options after decompression depend on the severity of the stenotic segment.

1. For mild to moderate stenosis without collaterals on static venogram, the patient can be left on anticoagulation. Most of such lesions remodel with time after bony decompression and venolysis.[27] Balloon angioplasty itself can cause further endothelial injury. For severe stenosis with multiple collaterals, immediate balloon venoplasty and even stenting can be considered. In patients undergoing angioplasty immediately after decompression, 100% patency at 4 years had been reported in a small case series.[28]
2. Patch venoplasty is reserved for patients with severe stenosis who are symptomatic despite bony decompression, external venolysis and balloon venoplasty.[29]
3. Symptomatic patients with total occlusion after thoracic outlet decompression can be managed with subclavian vein bypasses or jugular vein transposition (JVT). JVT is an excellent option for reconstruction of short occlusions limited to 5–6 cm.[30] For the longer occlusions, interposition grafting is a more practical option with good symptomatic improvement.[31]

Conclusion

Venous compression syndromes, such as PSS and MTS, are the common causes of DVT and post-thrombotic symptoms in the upper arm and lower limbs of young individuals, respectively. Early identification of these syndromes and appropriate treatment can decrease significant disability. Many aspects of their management are unclear. A review of the current evidence about these conditions is important for all vascular surgeons.

Conflict of Interest

None

Author Contributions

Study conception: ZUR
Investigation: ZUR
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Critical review and revision: ZUR
Final approval of the article: All authors
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