A Very Rare Etiology of Deep Vein Thrombosis 'KILT Syndrome'

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A Very Rare Etiology of Deep Vein Thrombosis- 'KILT Syndrome'

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Background

Deep vein thrombosis (DVT) is rare in young people and in most cases requires a very high index of suspicion. Anomalies of IVC account for 5% of DVT in patients under 30 years old[1, 2] Development of IVC occurs in the 4th to 8th week of gestation, and the complexity of its development poses opportunities for malformation to occur.[3]

Case

A 29 year old female presented with pain and diffuse swelling of her left lower extremity of 48 hour duration. She has history of oral contraceptive use since 2 years which was switched to Drospirenone and ethinyl estradiol combination 3 months ago. On admission, she was afebrile and her vital signs were stable. Physical exam showed extensive swelling of the left lower extremity with intact pulses and no skin changes. Venous duplex showed extensive occlusive thrombus in the left ilio - femoro – popliteal system. Thrombus was also identified in the infrarenal inferior vena cava (IVC) extending into the left common and external iliac vein (Figure 1, 2). On thrombophilia workup she had no prothrombin gene or Factor V Leiden mutation. She had low antithrombin III, protein S and protein C which was attributed to the consumption of coagulation factors and was appropriate in the setting of a large DVT. Anticardiolipin and beta-2-glycoprotein I antibodies were normal.

Patient underwent thrombolysis of the left ilio - femoral DVT and infrarenal caval thrombus. Post thrombolysis venogram showed severe stenosis at the junction of the left common iliac vein and IVC (Figure 3). IVC was diffusely irregular in contour suggesting chronic changes with a short segment of severe stenosis of the infrarenal IVC. There was a large paralumbar collateral which extended from the mid portion of the IVC on the left upto the diaphragm where it rejoined the IVC (Figure 4). There was no inflow seen into the IVC from the right common iliac vein suggesting flow through a duplicated IVC or a collateral. The stenosis at the junction of IVC and left common iliac vein was dilated followed by placement of a stent (16mm x 4mm). The short segment stenosis in the IVC just below the renal veins was also treated with balloon angioplasty. Retroperitoneal ultrasound showed hypoplastic left kidney measuring only 6.9 cm and enlarged right kidney measuring 14 cm (Figure 5 & 6). She was initiated on enoxaparin further bridged over to warfarin at the time of discharge.

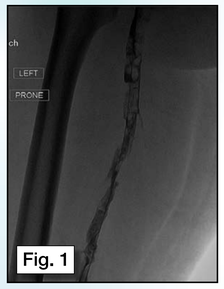

Fig. 1

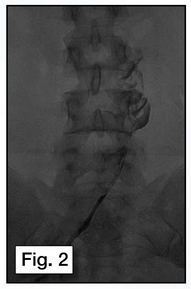
Fig. 2

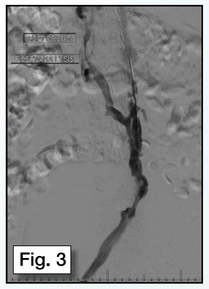
Fig. 3

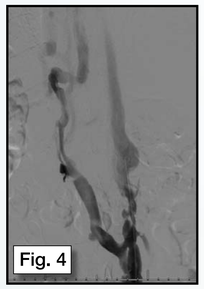
Fig. 4

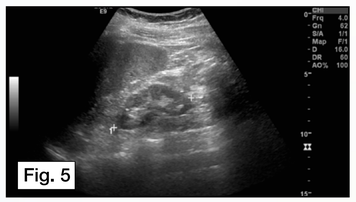
Fig. 5

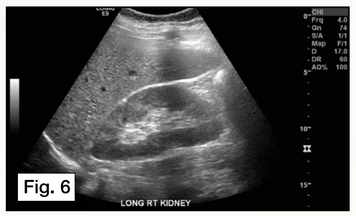

Fig. 6

Figure 1: Venogram showing thrombus in the left superficial femoral vein.
Figure 2: Venogram showing thrombus in the femoroiliac veins and IVC.
Figure 3: Post thrombolysis stenosis at the junction of the left common iliac vein and IVC.
Figure 4: Stenosis of the infrarenal IVC with paralumbar collateral.
Figure 5: Hypoplastic left kidney 6.9 cm.
Figure 6: Enlarged right kidney 14 cm.

Discussion

DVT occurs with a prevalence of 1 in 1000 adults. It is seen less in younger population with an estimated incidence of 1 in 10,000 (10 times less).[4, 5] IVC anomalies are an important and often overlooked risk factor for DVT in young patients. IVC anomaly and DVT in association with renal anomaly is very rarely reported in literature. Van Veen et al have proposed this striking association to be termed "KILT" syndrome (kidney anomaly, inferior vena cava anomaly, and leg thrombosis).[6]

There is literature evidence to suggest IVC abnormalities as an independent risk factor for DVT. Additional prothrombotic events could potentially increase the risk further.[7] In our patient the use of oral contraceptive pills was a potential trigger in otherwise silent anomalous IVC system. Venous thrombosis in young individuals should prompt work up to rule out venous anomalies in addition to hypercoagulable work up.

This case highlights the association of IVC anomalies with hypoplastic kidney as the knowledge about this could hold future clinical implications. Contralateral renal hypertrophy can compensate for the loss in kidney function, however this has far reaching implications especially when considering nephrotoxic medications or during future pregnancies. These patients should be monitored long term for proteinuria and early onset hypertension.

Learning Points

- 'KILT syndrome' is a very rare association of IVC and renal anomaly associated with deep vein thrombosis.
- Symptoms derived from these anomalies sometimes could be silent. Once diagnosed, these patients need long term follow up for proteinuria and early onset hypertension.
- Due to high thrombotic risk, one should consider avoiding risk factors including oral contraception and prolonged immobilization in these patient population.
- Rarity in literature would preclude definite conclusions, but consider prolonged if not life long anticoagulation for venous thrombosis in this patient group.

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