**KILT (Kidney and IVC Abnormalities with Leg Thrombosis) Syndrome in a 41-Years-Old Man with Loin Pain and Fever**

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**A R T I C L E   I N F O**

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**A B S T R A C T**

KILT syndrome is a rare condition composing the triad of kidney and inferior vena cava anomaly and extensive venous thrombosis. We present a case of newly diagnosed KILT syndrome in a 41-years-old gentleman presenting with loin pain and fever. Reviewing previous case reports, KILT syndrome is usually an incidental finding on imaging studies and there is a wide scope of initial clinical presentations. However, recent evidence suggests IVC anomaly may have caused subsequent renal hypoplasia. Identification of the underlying etiology may be helpful in planning early vascular intervention to treat the condition.

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**Introduction**

KILT syndrome — Kidney and IVC abnormalities with Leg Thrombosis — was first reported by Van Veen et al. With its low prevalence, there is currently limited knowledge about the condition. We present a case of a 41-years-old gentleman with newly diagnosed KILT syndrome presented with loin pain and low-grade fever. The renal and IVC anomalies were incidentally identified on the CT scan. While majority of the reported cases of KILT syndrome focus on the discussion of IVC anomalies, we try to look at the condition from a urological perspective.

**Case presentation**

A 41 years old male presented with first episode left loin pain and dysuria for 4 days. The pain radiated down to the suprapubic region, with intermittent dull aching nature. There was no hematuria, stone passage or gastrointestinal symptoms. He is a non-smoker non-drinker, with known well-controlled hypertension diagnosed in his late thirties during health check.

Low-grade fever was noted upon admission. Physical exam was unremarkable. Bedside ultrasound showed a prominent right kidney but the left kidney was barely visualized. Blood count and renal function were both normal.

Subsequently a computed tomography (CT) scan was arranged, showing extensive deep venous thrombosis (DVT) involving bilateral common iliac veins, left common and external femoral veins, with small filling defects over the right common and external femoral veins. The infrarenal portion of the IVC has a notably small calibre of 0.8 cm (Fig. 1). Left kidney is hypoplastic, with compensatory hypertrophy of the right kidney.

Further physical exam revealed mild left calf swelling with insignificant circumferential difference. Typical findings of chronic DVT including tortuous veins, skin hyperpigmentation or venous ulcers were absent. Doppler ultrasound confirmed the diagnosis of extensive left sided DVT extending from the common femoral vein to the popliteal vein (Figs. 2 and 3).

He was started on low-molecular weight heparin injection and referred to the cardiothoracic team for assessment. Eventually the calf swelling and fever both subsided with medical treatment. He was followed up by the hematologists and vascular surgeons and planned for lifelong anticoagulation therapy.

**Discussion**

IVC anomaly is a rare congenital condition with an estimated prevalence of 0.005% to 1% of the general population. In a study performed by Sagban et al., right and left kidney
anomalies were identified in 6% and 2.7% of IVC anomaly cases respectively.

The presentation of acute DVT appears quite diverse. Gayer et al reviewed the clinical presentation of 11 cases of known IVC anomaly with acute iliac or femoral venous thrombosis on CT. Three of them presented as loin pain, hematuria and low back pain. Others presented typically with lower limb swelling. And the patient presented with low back pain is the only individual with KILT syndrome. Another patient who presented with loin pain and hematuria has normal bilateral kidneys on the CT scan. Despite the small population size, we notice that DVT presentation can be quite variable, and loin pain during DVT attacks is possible and does not necessarily suggest underlying KILT syndrome.

In 2015, Duicu et al presented the first known case of siblings suffering from KILT syndrome. The 12-years-old elder brother presented with acute abdomen and an emergency appendectomy was performed. Intra-operatively, the surgeons noted a large right kidney. CT shows an enlarged right kidney with absent right renal vein flow, and the IVC segment from the confluence of hepatic veins to the common iliac veins is absent. The left kidney appears normal otherwise. Follow up magnetic resonance imaging (MRI) 1 year later demonstrates a hypoplastic right kidney. His younger sister presented with acute onset right abdominal and flank pain followed by lower limb pain at the age of 12. Bedside ultrasound confirms the diagnosis of venous thrombosis. CT scan again demonstrates the absence of the supra- and infra-renal segments of IVC and a hypoplastic left kidney. Extensive investigations including thrombotic tendencies, clotting factors screening, autoantibodies detection and selected genetic analysis were arranged but no abnormalities were detected. They have a younger third sibling but her IVC and kidneys are normal.

The brother’s presentation brings new insight into the chronological relationship of IVC and renal anomaly. As reflected by his follow up MRI scan a year after, renal hypoplasia may be a consequence of venous malformation. This is clinically significant as renal hypoplasia may be reversible with early identification of IVC anomaly. Early vascular intervention or reconstruction may theoretically prevent the development of renal hypoplasia. However, complete renal agenesis on presentation is also reported previously, possibly representing another group of variant when renal anomaly happens congenitally. This requires a more comprehensive review.

The sibling presentation also directs future investigations along any underlying genetic components of IVC anomalies. Duicu et al tried to investigate along currently known hematological conditions, such as factor V Leiden and prothrombin gene 20210A.
Successful genetic identification allows selection of high-risk cases with early surgical planning to restore the IVC anomaly if present.

**Conclusion**

KILT syndrome is commonly an incidental finding during venous thrombosis or acute loin pain. No specific clinic presentations have been identified and the renal biochemical function tests may appear normal. Apart from venous thrombosis, we should also keep an active eye on medical conditions such as young onset hypertension or renal function derangement as a result of renal hypoplasia. Long-term follow up studies of patients with KILT syndrome may be helpful to unveil associated co-morbidities.

Future investigation should target on the underlying etiology for early detection. Possible directions may include embryological perspectives of renal and IVC genesis or genetics studies. If renal hypoplasia is caused by venous abnormalities, we should work on the feasibility of prevention by early vascular intervention.

**Conflicts of interest**

The authors declare that there is no conflict of interest.

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