Kidney and Inferior vena cava abnormalities and Leg Thrombosis (KILT) syndrome in a young man mimicking retroperitoneal lymphoma

Authors: Małgorzata Twardowska-Kawalec, Monika Wieliczko, Joanna Matuszkiewicz-Rowińska, Krzysztof Milczarek, Jolanta Małyszko

Article type: Clinical image

Received: October 28, 2019.

Accepted: November 29, 2019.

Published online: December 4, 2019.

ISSN: 1897-9483
Kidney and Inferior vena cava abnormalities and Leg Thrombosis (KILT) syndrome in a young man mimicking retroperitoneal lymphoma

Małgorzata Twardowska-Kawalec\textsuperscript{1}, Monika Wieliczko\textsuperscript{1}, Joanna Matuszkiewicz-Rowińska\textsuperscript{1}, Krzysztof Milczarek\textsuperscript{2}, Jolanta Małyszko\textsuperscript{1},

\textsuperscript{1}Department of Nephrology, Dialysis and Internal Diseases, Medical University of Warsaw, Warsaw, Poland
\textsuperscript{2}Department of Interventional Radiology, Medical University of Warsaw, Warsaw, Poland

Correspondence to:
Prof Jolanta Malyszko, Department of Nephrology, Dialysis and Internal Diseases, Warsaw Medical University, tel. +48 22 599 2660, mail: nefrologia@wum.edu.pl, jolmal@poczta.onet.pl

Short title: KILT syndrome mimicking lymphoma

Conflict of interest: none declared

**Key words**: collateral venous abdomen circulation, deep venous thrombosis (DVT), inferior vena cava (IVC) occlusion, magnetic resonance imaging, retroperitoneal mass

**Clinical image:**
A 34-years-old previously healthy man, was referred to the hospital due to acute edema of the left leg caused by deep vein thrombosis-DVT and high suspicion of lymphoma.

A few weeks before the admission, he started intensive physical activity for weight reduction, then he suffered from gastrointestinal infection with vomiting and diarrhea leading to dehydration. Leg edema with fever developed shortly afterwards. Ambulatory
sonography and computed tomography-CT performed without the contrast showed occlusion of inferior vena cava-VCI and retroperitoneal packets of enlarged lymph nodes [Fig.1- A]. On admission he was in a stable clinical condition with fever 38°C, left leg, lumbar region pain while moving. Edema and redness of the left leg with positive Homman’s sight were found on physical examination without palpable peripheral lymph nodes. Laboratory tests showed elevated C-Reactive Protein-2219nmoL/L (normal range 0.76-28.5 nmol/L), procalcitonin-1.4ng/L (normal <0.05μg/L), fibrinogen-9.52 g/L (range normal 1.5-4.5g/L), D-dimer-7287 μg/L (normal <460 μg/L), platelet count-437x10⁹/L (normal range 150-350x10⁹/L), creatinine-120 μmol/L (normal range 53-106 μmol/L), whereas lactic dehydrogenase and coagulation factors were normal. Testicular cancer was excluded as ultrasound of the testicles was normal. Low molecular weight heparin-enoxaparin was continued, and antibiotic (amoxycillin and clavulanic acid) was started. Biopsy of the enlarge retroperitoneal nodes under a tomography was performed however, sample for pathology assessment was insufficient and nonspecific and additionally seemed to be a clotted blood. To exclude a puncture of an aneurysm the CT scan with contrast and Doppler ultrasound were performed ad hoc, both showed surprisingly well-developed collateral venous abdomen circulation and confirmed once again the occlusion of VCI. Then magnetic resonance imaging-MRI and magnetic resonance angiography-MRA [Fig. 1-B-D] was performed showing occlusion of infrarenal part of VCI, alternative enormously developed, tortuous, partially baggy vertebrolumbar channels which were also clotted, thrombosis of both iliac veins with a greater thrombosis on the left side, hypotrophic left kidney and dilated azygos vein. An extended coagulogram, antinuclear antibodies-ANA, anti-neutrophil cytoplasmic antibodies-ANCA, anticardiolipin and anti-beta2glicoprotein antibodies were negative as well as cancer screening. The histopathology of “suspected lymph nodes” were described as a hyphae of crushed illegible connective tissues. Creatinine and inflammatory parameters normalized during
hospitalization. Enoxaparin and acetylsalicylic acid were recommended to continue for three months, the Doppler ultrasound was suggested as a method to evaluate the permeability of collateral venous circulation and legs veins. He was referred to hematology and angiology service for further diagnosis, treatment and surveillance. Acute lower extremity deep venous thrombosis in young adults is usually related to thrombophilia, immobility, trauma, surgery or malignancy [1]. Congenital occlusion of VCI is very rare and is usually find during the fetus development [2] and most patients are asymptomatic [3]. The vertebrolumbar route is involved in 95% of cases [2]. This rare congenital anomaly may mimic retroperitoneal lymphadenopathy [3,4], especially on CT without contrast [5], thus MRI-contrast enhanced is recommended [3]. The association between absence of VCI, renal abnormalities (mainly right kidney hypotrophy) and leg thrombosis has been termed Kidney and IVC abnormalities and Leg Thrombosis (KILT) syndrome [3]. Additional risk factors for thrombosis such as smoking, hormonal contraceptives, strenuous physical activity should be avoided [1]. It is important to recognize this entity to avoid unnecessary laparotomies and biopsies [2].

References


**Figure 1.**

A – computed tomography scan without contrast enhancement showing “suspected retroperitoneal mass”, red lines shows the borders of suspicious mass.

B – magnetic resonance, T1 sequence (LAVA), axial section - red arrows indicates multiple occluded tortuous vessels of collateral abdomen circulation mimicking enlarged lymph nodes. Enormously developed, tortuous, partially baggy vertebrolumbar channels as an alternative route in congenital inferior vena cava occlusion:

C – magnetic resonance angiography, T1 sequence, coronal section (TRICKS);

D – magnetic resonance angiography, venous phase, coronal section. venous phase, coronal section; red arrows on both scans shows enormously developed, tortuous, partially baggy vertebrolumbar channels and occluded internal left iliac vein (red star)