

Inferior vena cava agenesis presenting as deep venous thrombosis

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Background:

Inferior vena cava agenesis (IVCA) is a rare congenital malformation caused by dysgenesis during embryogenesis. The condition is often asymptomatic due to the development of collateral venous circulation. It carries a significant risk for deep venous thrombosis (DVT), especially in young people as a result of venous stasis in the lower extremities. The rarest form is infrarenal and renal segment hypoplasia, followed by hypoplasia only of the infrarenal segment. The first episode of deep venous thrombosis usually occurs before the fourth decade, with no gender predilection.

Case presentation:

A 17-year-old, previously healthy male presented to the emergency department with complaints of month-long pain in the right lumbar region and the right leg, following a punch during a hockey match. On the physical examination, there were signs of right leg swelling and skin discoloration. Laboratory tests revealed elevated D-dimer levels (5 mg/L). Color Doppler ultrasonography (CD US) and computed tomography were performed, revealing extensive venous thrombosis of both legs and pelvis. The infrarenal segment of the vena cava inferior (VCI) was missing. Instead, collateral pathways developed through paravertebral and lumbar veins, draining into a prominent azygos vein. Therefore, a treatment with low molecular weight heparin (LMWH) was immediately started. Following phlebography, pharmacomechanical thrombolysis was performed (alteplase and heparin infusion with clot aspiration and percutaneous transluminal angioplasty), achieving complete reperfusion. Two days after admission, the patient was discharged symptom-free and in good general condition with lifelong anticoagulation prophylaxis.

Conclusion:

IVCA is a highly underrecognized cause of DVT in the young population. Because of the condition's rarity, there are no specific guidelines for the management. Currently, treatment is mainly conservative and focused on preventing clot formation and recurrence (lifelong anticoagulation therapy, compression stockings). Further studies need to provide an optimal approach to patient management.