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INTERMEDIATE

CASE REPORT: CLINICAL CASE

Venous Thromboembolism in a Young Man With Fused Renal Ectopia

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ABSTRACT

A 19-year-old man presented with chest pain after a trans-Pacific flight. Venous thromboembolism was diagnosed and treated with catheter-directed thrombolysis. Genetic testing revealed factor V Leiden mutation. In addition to the flight history and genetic hypercoagulability, a renal abnormality causing an external compression over the inferior vena cava was suspected to be a contributing factor. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:2479–83) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 19-year-old White man presented with chest pain and left calf edema that had developed in the course of 6 days. He was an U.S. student studying in Japan who had recently come back from a temporary return to the United States. The trans-Pacific flight occurred 2 weeks before his hospital visit. His vital signs were stable. He expressed general back discomfort, as well as sharp right chest pain on inspiration. His left leg was swollen, with tenderness on dorsiflexion.

LEARNING OBJECTIVES

- Even when a genetic thrombophilia is identified, other contributing predispositions may also be present.
- Congenital renal abnormalities may be a causative factor in VTE development.
- Especially in young patients, thorough imaging evaluation is essential to identify rare anatomic causes of thrombosis.

PAST MEDICAL HISTORY

The patient had no past medical history of relevance, with only a previous diagnosis of a renal abnormality.

DIFFERENTIAL DIAGNOSIS

Acute chest pain after a long flight is a classic presentation of pulmonary embolism (PE).

INVESTIGATIONS

Laboratory tests showed an elevated D-dimer value. Results of other blood tests, including cardiac troponin and brain natriuretic peptide, were normal. Results of anticardiolipin antibody and lupus anticoagulant testing were negative. The 12-lead electrocardiogram showed normal sinus rhythm without ST-T wave changes. Transthoracic echocardiography showed normal contraction, and signs of right ventricular dysfunction were absent.

Contrast-enhanced computed tomography (CT) confirmed PE in the right pulmonary artery branches A8 to 10 and a large thrombus extending from the inferior vena cava (IVC) to the left femoral vein

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

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ABBREVIATIONS AND ACRONYMS

CDT = catheter-directed thrombolysis

CT = computed tomography

DVT = deep vein thrombosis

IVC = inferior vena cava

PE = pulmonary embolism

VTE = venous thromboembolism

(Figures 1A to 1C). Moreover, a fused ectopic kidney was discovered on his right side (Figure 1D). The proximal end of the thrombus was within the same transverse plane as the abnormal vessel (Figure 2). An ultrasound scan of the lower extremity veins demonstrated areas of noncompressibility consistent with the CT findings. With the patient's consent, additional screening for inherited thrombotic disorders was conducted. He tested positive for factor V Leiden mutation.

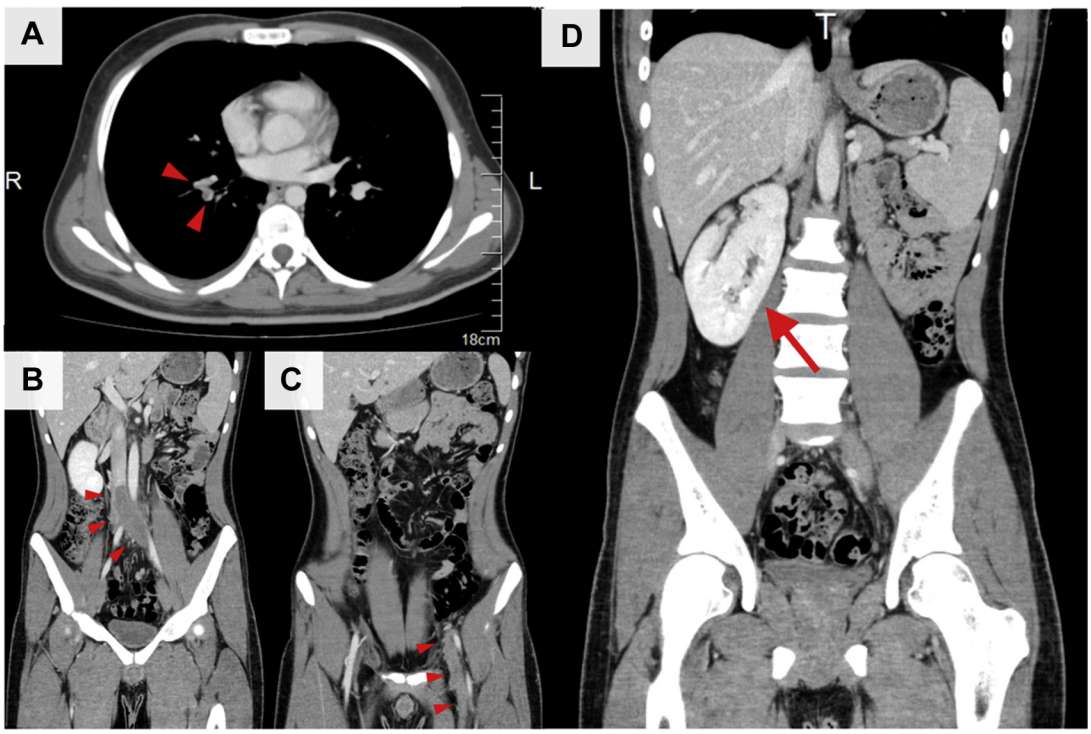
MANAGEMENT

The patient was promptly started on anticoagulant therapy with heparin. Despite dosage escalation for 5 consecutive days, a follow-up CT scan revealed an increase in the thrombus size. His symptoms persisted. Given his poor response to anticoagulation, we placed a temporary IVC filter and started systemic urokinase administration. Eight days later, the patient experienced increasing pain in both of his extremities, and there was still no significant change in clot size. Subsequently, we conducted pharmacomechanical catheter-directed thrombolysis (CDT). Suction thrombectomy and balloon angioplasty were performed, followed by continuous infusion of urokinase through a multiple-side-hole catheter. After another 8 days, a venous angiogram revealed patency of the affected veins and partial reduction in clot size (Figure 3). The D-dimer level, once reaching 58.9 μg/ml, dropped to 2.0 μg/ml. His symptoms also improved remarkably. With the proximal part of the residual clot still remaining, the filter, initially planned to be retrieved, was left in place. On hospital day 35, the patient was finally discharged with oral anticoagulant therapy.

DISCUSSION

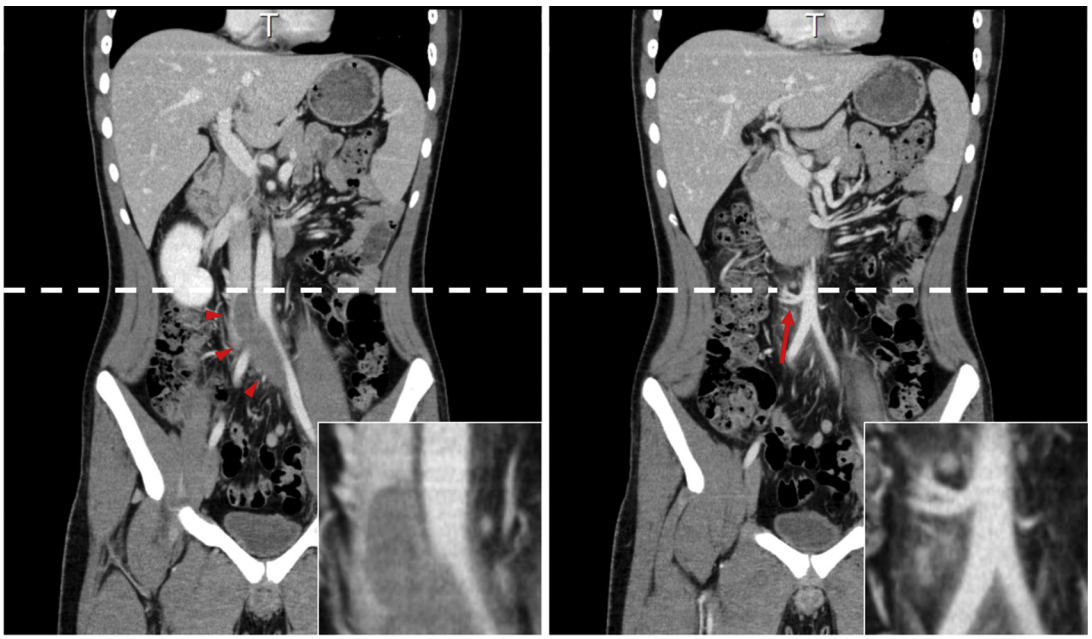
In stable patients with PE without right ventricular dysfunction, thrombolysis is not routinely recommended because of bleeding risks. Nonetheless, there

FIGURE 1 Contrast-Enhanced Computed Tomography



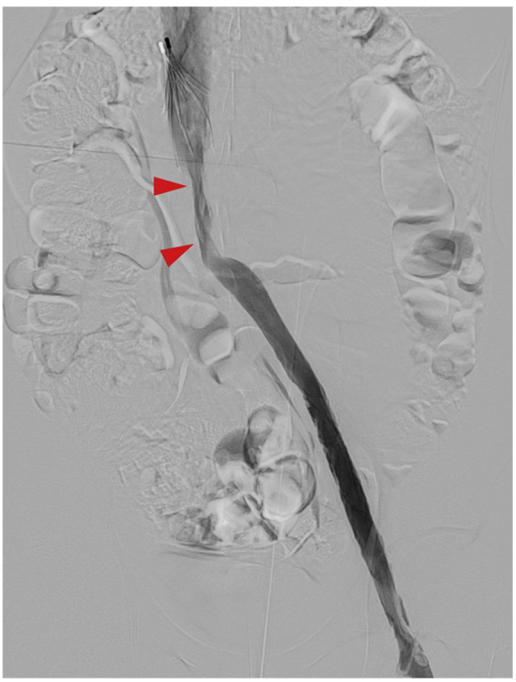
(A) Filling defects in the pulmonary vasculature confirmed the diagnosis of pulmonary embolism **(arrowheads). (B and C)** A large venous thrombus extended from the inferior vena cava to the femoral vein **(arrowheads). (D)** A unilateral crossed-fused ectopic kidney **(arrow)** was discovered on the right side. L = left; R = right.

FIGURE 2 Contrast-Enhanced Computed Tomography



The **dotted white line** shows the level of the cranial end of the thrombus **(arrowheads),** matching that of the accessory renal artery **(arrow).**

FIGURE 3 Post-Treatment Venous Angiography



Post-treatment venous angiography revealed patency of the iliofemoral vein and the inferior vena cava. The proximal remnant of the thrombus **(arrowheads)** remained in the inferior vena cava.

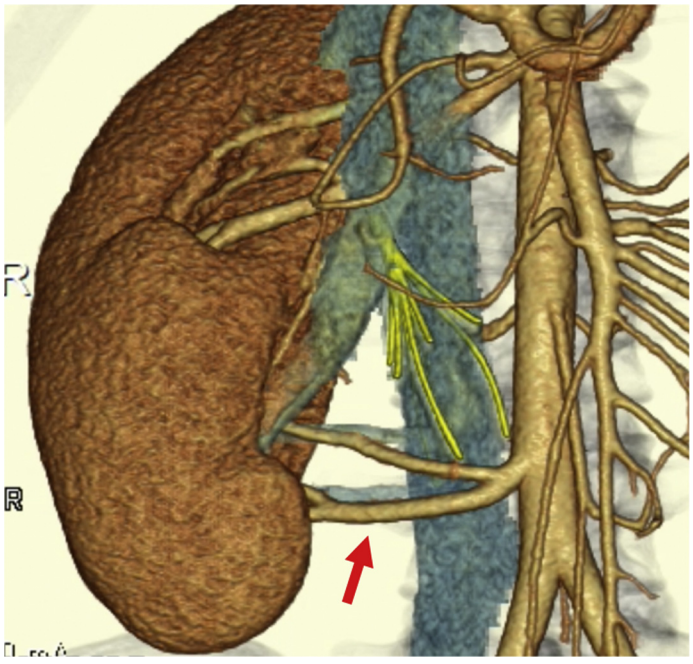
is abundant evidence supporting pharmacomechanical CDT to manage PE in patients with proximal deep vein thrombosis (DVT). CDT is expected to reduce the incidence and severity of post-thrombotic syndrome, a common complication of proximal DVT (1,2). Studies have shown the safety and effectiveness of CDT even during pregnancy and in patients with cancer (3,4). After an inadequate response to anticoagulation and systemic thrombolysis, our patient with no comorbidity was a good candidate for CDT.

Temporal IVC filter insertion in patients with iliocaval DVT and anticoagulant therapy failure is supported in multiple guidelines. However, complications of prolonged dwell time and filter nonretrieval have become major issues in recent years (5). We deemed retrieval before discharge to be unsafe in our case because of the persistent risk of significant PE from residual IVC thrombus. Accordingly, regular follow-up visits were planned to arrange for timely retrieval of the filter.

Two clear contributors to thrombosis in this patient were the long flight and the factor V Leiden mutation. The refractory nature of the thrombus in our case can be explained by the partial organization that it had undergone during the first post-flight week.

The renal abnormality was a unique feature in this patient. The proximal end of the thrombus was

FIGURE 4 3-Dimensional Reconstructed Computed Tomography



The fused ectopic kidney and an accessory artery **(arrow).** The artery, overlying the inferior vena cava, was suspected to represent an underlying predisposition for thrombosis. R = right.

exactly at the same level as the abnormal renal artery, thus raising suspicion that the abnormality had caused stasis of IVC flow. Although the right renal artery runs posteriorly to the IVC in normal subjects, our patient had an accessory artery that coursed anteriorly, positioning the IVC between the iliac spine and the artery itself (**Figure 4**). Such anatomic arrangement is comparable to that of May-Thurner syndrome, a well-known condition in which thrombosis in the left iliac vein results from compression between the right iliac artery and the iliac spine. In young patients, multiple cases of thrombosis secondary to anatomic abnormalities, such as IVC malformations and May-Thurner syndrome, have previously been described (6,7). To our knowledge, only 1 case of DVT caused by a renal abnormality has been reported in an adolescent patient (8). In factor V Leiden carriers younger than 60 years of age, the incidence of venous thromboembolism (VTE) does not differ significantly from that in noncarriers (9). With more than one-half of patients with VTE carrying 3 or more risk factors for thrombosis (10), anatomic abnormalities may be frequently missed predispositions.

However, evidence to support our hypothesis is limited. An abdominal duplex ultrasound scan during follow-up showed no obvious signs of IVC stenosis or restriction in IVC flow. Further testing, such as intravascular ultrasound, was suspended because the patient relocated to the United States. The renal ectopy may have been merely an incidental finding unrelated to thrombosis. Another possibility is that the aberrant artery had trapped a pre-formed iliofemoral thrombus and acted as an "inborn IVC filter," thereby preventing the thrombus from traveling further caudally. We could not reach a definite conclusion about the relationship between VTE development and renal abnormality in this patient.

FOLLOW-UP

Follow-up CT 5 months after discharge finally showed dissolution of the thrombus. With his study abroad program in Japan coming to a close, the patient wished to discuss filter retrieval with the primary care physician in his home country. The patient has experienced no symptoms of post-thrombotic syndrome and no complications of anticoagulation.

CONCLUSIONS

The arterial abnormality associated with renal ectopia, in addition to the patient's genetic hypercoagulability and limited mobility from a long flight, may have contributed to thrombus formation. When our hypothesis is confirmed, further treatments such as stenting should be considered to avoid relapse.

AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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