A rare case of pulmonary embolism in a patient with interrupted inferior vena cava with azygous continuation and polysplenia

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Background

- Anomalies of the inferior vena cava (AIVCs) are typically asymptomatic and often associated with congenital heart disease.
- Such anomalies are becoming more commonly detected and have a reported prevalence of 0.6%.
- Despite this, the presence of pulmonary embolism (PE) in this population is underreported.

Patient Presentation

History of Present Illness:

- 62-year-old otherwise healthy female presented to the emergency department with a complaint of shortness of breath following a 22-hour flight two days prior to presentation
- Associated chest pain, dyspnea on exertion, and non-productive cough
- No history of pulmonary disease, stroke, bleeding disorder, tobacco use, and sick contacts

Physical Exam:

- Vital signs: normotensive, RR 28 breaths/min, HR 93 bpm, SpO2 95% RA
- Exam: no acute distress, unremarkable exam

Laboratory Data:

- CBC and CMP normal
- High sensitivity troponin-I 311 ng/L, BNP 345 pg/mL, CRP 42 mg/L, IL-6 44.6 pg/mL, SARS-CoV-2 positive

Diagnostics:

- EKG: sinus rhythm without significant ST or T wave changes
- Chest x-ray: No airspace disease, prominent azygous vein
- CTA: Bilateral PEs with saddle embolism and straightening of IVS with increased RV/LV ratio, dilated azygous vein, contrast reflux in hepatic IVC, polysplenia
- Lower extremity US: occlusive DVTs in the left distal femoral vein and popliteal vein

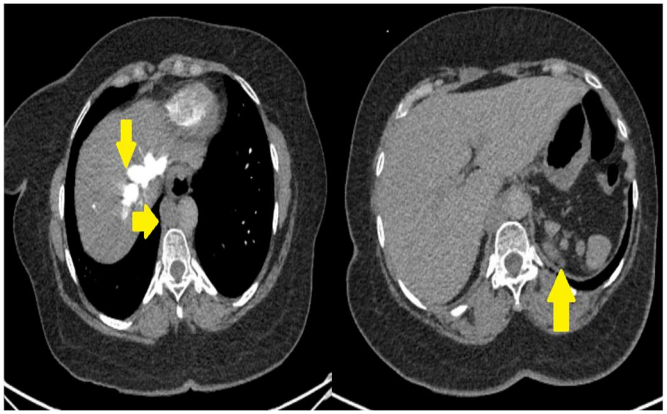

Figure 1 (left): Axial view of the CTPE showing the IVC drain into hepatic vein (top) and enlarged azygous vein adjacent to the descending aorta (bottom).
Figure 2 (right): Axial view of the CTPE demonstrating fragmented spleen consistent with polysplenia.

Clinical Course

- A diagnosis of high risk submassive PE was made and high intensity heparin therapy was initiated.
- PERT consulted and the decision was made to pursue mechanical thrombectomy.
- Right heart catheterization was unable to be performed because the equipment was unable to be advanced to the right heart via the femoral approach.
- A review of the imaging was conducted intraprocedurally, and angiography suggested the catheter was in the azygous vein and this made a connection with the superior vena cava.
- The procedure was aborted, and the patient was continued on therapeutic anticoagulation.
- The patient required supplemental oxygen as coronavirus disease progressed, but was discharged on lifelong DOAC after 2 –week hospital stay.

Discussion

- Pulmonary embolism with concurrent anomalous IVC is exceptionally rare; however, the recognition of such an abnormality prior to invasive procedures is essential for the interventionalist.
- Significant dilation of the azygous vein, more so than commonly seen in congestive heart failure, can be a clue in the detection of this abnormality.
- Additionally, polysplenia can be an indication of an IVC anomaly.
- Dedicated imaging of the abdominal vasculature may also be helpful prior to proceeding with an intervention if an anomalous IVC is suspected.
- Anomalies of the IVC can alter both the type of intervention and the access site. Interventions requiring large bore catheters, such as a 24 French, can be challenging in the setting of an anomalous IVC, as these procedures are primarily done via femoral access.
- In the event of an anomalous IVC, an internal jugular (IJ) approach would be required, and early recognition would avoid any unnecessary attempt at femoral access.

References

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