Disseminated intravascular coagulopathy secondary to type I endoleak case report: challenges in diagnosing and managing overlapping symptoms in end-stage renal disease

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Research Article

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Abstract

Introduction: Endoleak-associated DIC is an understudied condition with potentially overlapping symptoms that can increase difficulty of diagnosing and managing patients, especially in patients with comorbid conditions such as renal failure.

Case report: A 79-year-old female with significant past medical and surgical histories of stage V chronic kidney disease and abdominal aortic and right external iliac artery aneurysms status-post endovascular presented to an academic hospital for urgent dialysis catheter placement due to end-stage renal disease (ESRD) with symptoms of hypervolemia, electrolyte abnormalities and pancytopenia. Nephrology was consulted on admission. Temporary catheter was placed without complication however patient continued to bleed from that site. Initially, there was suspicion for uremic thrombocytopenia; however, despite tolerating multiple dialysis sessions, patient continued to bleed even after receiving platelet transfusions. Vascular surgery was unable to stop the bleeding. Additional workup revealed abnormal coagulation panel, including low fibrinogen. Hematology was consulted, concerning for possible malignancy, immune thrombocytopenic purpura, or disseminated intravascular coagulopathy (DIC). CT abdomen revealed type I endoleak (6.5cm in diameter) at proximal bilateral common iliac arteries. Cryoprecipitate was given due to low fibrinogen, secondary to endoleak-associated DIC. The bleeding subsequently stopped and patient was discharged with appropriate outpatient appointments including dialysis and vascular surgery.

Conclusion: which has overlapping symptoms with many diseases in ESRD patients.

Introduction

Diagnosing and treating etiologies of pancytopenia can be difficult in end-stage renal disease (ESRD) patients given their comorbid risks. Given that ESRD is known to be a major cause, frequently patients undergo dialysis sessions with many subsequently improving [1]. Endoleaks after aneurysm repair should be a differential in the appropriate clinical context. Disseminated intravascular coagulation (DIC) is a rare complication of endoleaks. There is a paucity of research and clinical data regarding DIC due to endoleaks. Our case illustrates the clinical dilemmas in diagnosing, treating and managing overlapping symptoms of this condition.

Case Report

A 79-year-old female with past medical and surgical histories of stage V chronic kidney disease (CKD), hypertension, heart failure with preserved ejection fraction of 67%, abdominal aortic and right external iliac artery aneurysms status-post endovascular repaired 7/2020 and 11/2020 (EVAR) respectively presented to an academic hospital in United States for management of hypervolemia due to CKD requiring urgent right internal jugular vein (RIJV) catheter placement and hemodialysis. Patient used to be active at baseline, able to perform activities of daily living, but became more sedentary during the past year due to experiencing worsening bilateral lower extremity swelling and exertional dyspnea. Outpatient

nephrology has been following patient who was refusing dialysis prior to this admission; however, her worsening symptoms changed her mind.

When patient was initially hospitalized, she was found to have pancytopenia with white blood count (WBC), hemoglobin (Hgb), and platelet count showing 3.0 x 10^3/uL, 8.9g/ dL (normocytic), and 76 x 10^3/uL respectively. Nephrology and primary team believed low Hgb and platelet could be explained by anemia of chronic kidney disease and uremic thrombocytopenia. Blood urea nitrogen (BUN) and creatinine (Cr) on admission were 98 and 7.32 respectively. Sepsis was low on the differential since patient did not demonstrate other associated symptoms.

Due to patient's deteriorating condition, decision was made to urgently place RIJV catheter to begin dialysis, while allowing primary team to begin pancytopenia work-up without delaying critical therapy. Primary team spoke with patient's outpatient primary care physician who was also a hematologist and learned that at her previous hospitalization one year ago at another institution, there was concern for possible heparin-induced thrombocytopenia but patient was never tested. Subsequently, she was tested and found to have negative heparin-induced thrombocytopenia platelet factor 4 antibody and negative serotonin-release assay on this admission.

Patient was able to tolerate dialysis but had continuous bleeding at the catheter site. Initially, vascular surgery and nephrology believed additional dialysis sessions will improve her platelet count and therefore stop the bleeding. However, she continued to bleed despite receiving more than four dialysis sessions. Vascular surgery was at patient's bedside daily to change her dressing and suture her site, which did not stop the bleeding. Meanwhile, hematology was consulted regarding her pancytopenia. Possible malignancy work-up was initiated including leukemia, lymphoma, and immune thrombocytopenic purpura (ITP) as well as DICs. Patient was also started on empiric prednisone to cover for possible ITP although it was low on the differential.s

Computed tomography (CT) of abdomen and pelvis found type I endoleak (6.5cm in diameter) at proximal bilateral common iliac arteries (see Image section, Fig. 1). Patient's coagulation panel was obtained which showed low fibrinogen (94 mg/dL) and d-dimer greater than the upper limit of the test value (> 10,000 ng/mL). Thromboelastogram showed low max amplitude (< 40 mm), low fibrinogen function, low adenosine-5'-diphosphate, low kaolin with heparinase, normal activator F and normal R value, suggesting platelet dysfunction and possible DIC. In addition to having possible dialysis-associated thrombocytopenia, it was theorized that patient may be experiencing chronic DIC secondary to endoleak and required surgical assessment.

Vascular surgery contacted patient's surgeon at a different institution who performed her endovascular repairs. Immediate outpatient follow-up was recommended once patient was stable to be discharged.

To stabilize patient from blood loss and thrombocytopenia, she was given a total of 2 units of packed red blood cells, 6 units of platelet and 2 rounds of desmopressin during her hospitalization. She was also found to have low fibrinogen and was given 2 units of cryoprecipitate. The combination of

anticoagulation used during dialysis, platelets and antifibrinolytic therapies stopped the bleeding. After hemodynamic stabilization, she was discharged after outpatient surgical follow-up and hemodialysis appointments were arranged.

Discussion

We presented a case of a patient with multiple comorbid risks for pancytopenia who had continuous bleeding at her RIJV catheter site, complicating her clinical work-up and hospital course. We hypothesized that patient might have been experiencing chronic DIC after EVAR secondary to type I endoleak.

Studies have shown that up to 4% of patients with rupture or dissection of aortic aneurysms may have DIC, but DIC secondary to endoleaks, including type I, after EVAR is rarely reported with a paucity of research [2, 3]. DIC is a syndrome leading to inappropriate activation of hemostasis, thrombosis, and consumption of coagulation factors and platelets leading to increased risks for thromboembolism and bleeding. In the beginning, the constant endoleak to the aneurysm sac increased exposure of denuded endothelium and release of tissue factors, resulting in clotting factor consumption [4]. The turbulent blood flow can cause intravascular hemolysis of red blood cells through shear stress, further activating coagulation cascade and leading to DIC [5]. Although rare, endoleaks should be on the differential for any patient who has EVAR with thrombocytopenia as this could be life threatening with few medical interventions available. Work-up requires elucidating the underlying etiology with involving specialists and imaging to rule out endoleaks.

Our case illustrates the difficult nature in diagnosing endoleak-associated DIC, particularly in patients with comorbid risk factors such as uremic thrombocytopenia and anemia of chronic kidney disease. This is one of the few case reports discussing thrombocytopenia diagnosis and management in patients with endoleak and ESRD.

Conclusion

Our case demonstrates the difficult nature in diagnosing and managing dialysis patients with concern for endoleaks and refractory thrombocytopenia. Although disseminated intravascular coagulation due to endoleaks from surgically-repaired aneurysms is rare, it can be life-threatening and needs to be on the differential for patients with surgical history experiencing refractory thrombocytopenia. Urgent recognition of this the condition can improve outcomes and save additional limited hospital resources, including units of blood and platelets.

Declarations

1. Funding: Not applicable
2. Conflicts of interest/Competing interests: The authors declare that there is no conflict of interest regarding the publication of this article.

3. Ethics approval: Ethical approval is not required at our institution to publish an anonymous case report.
4. Consent to participate: Not applicable
5. Written consent for publication: Report has been anonymized and written consent was obtained.
6. Availability of data and material: Not applicable
7. Code availability: Not applicable
8. Authors' contributions: All authors of this manuscript wrote the manuscript, managed and treated the patient.

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Figures

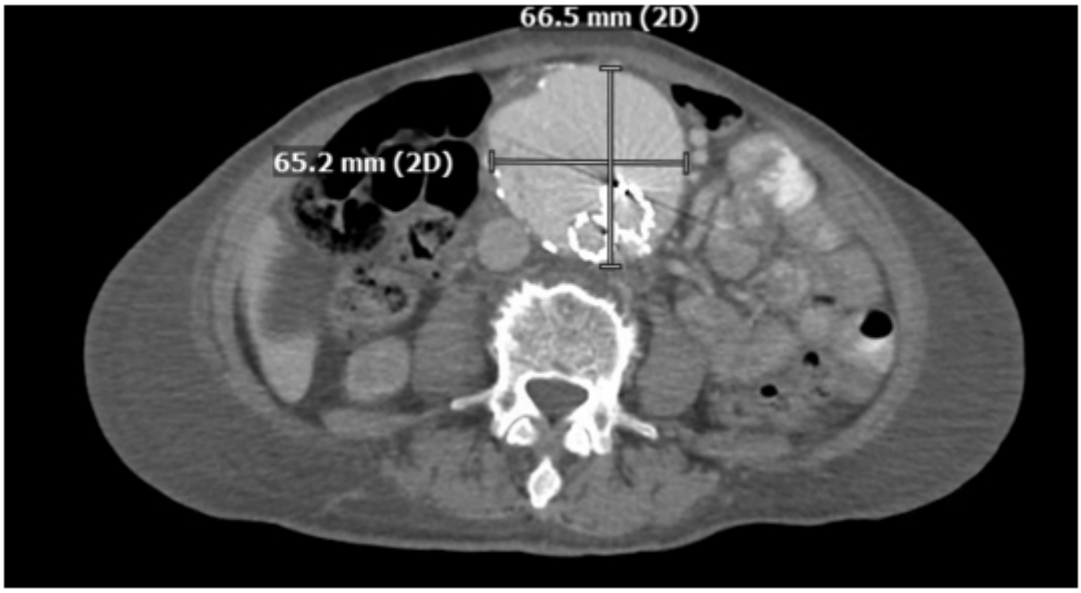

Figure 1

Media type I endoleak (6.5cm in diameter) at proximal bilateral common iliac arteries see on CT of abdomen and pelvis

Supplementary Files

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