Budd-Chiari syndrome secondary to catheter-associated inferior vena cava thrombosis

Síndrome de Budd-Chiari secundária a trombose de veia cava inferior associada a cateter

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Abstract

Introduction: Patients with chronic kidney disease (CKD) are at increased risk for thrombotic complications. The use of central venous catheters as dialysis vascular access additionally increases this risk. We describe the first case of Budd-Chiari syndrome (BCS) secondary to central venous catheter misplacement in a patient with CKD. **Case report:** A 30-year-old female patient with HIV/AIDS and CKD on hemodialysis was admitted to the emergency room for complaints of fever, prostration, and headache in the last six days. She had a tunneled dialysis catheter placed at the left jugular vein. The diagnosis of BCS was established by abdominal computed tomography that showed a partial thrombus within the inferior vena cava which extended from the right atrium to medium hepatic vein, and continuing along the left hepatic vein. Patient was treated with anticoagulants and discharged asymptomatic. **Discussion:** Budd-Chiari syndrome is a rare medical condition caused by hepatic veins thrombosis. It can involve one, two, or all three of the major hepatic veins. It is usually related to myeloproliferative disorders, malignancy and hypercoagulable states. This case calls attention for inadvertent catheter tip placement into hepatic vein leading to this rare complication. **Conclusion:** Assessment of catheter dialysis tip location with radiological image seems to be a prudent measure after each procedure even if the tunneled dialysis catheter has been introduced with fluoroscopy image.

Keywords: Budd-Chiari syndrome; central venous catheters; venous thrombosis.

Resumo

Introdução: Pacientes com doença renal crônica (DRC) apresentam risco aumentado de complicações trombóticas e o uso de cateter venoso central para realização de hemodiálise aumenta este risco. Nós descrevemos um caso de síndrome de Budd-Chiari (SBC) causado pelo mal posicionamento de um cateter de diálise em um paciente com DRC e, para nosso conhecimento, este é o primeiro caso relatado na literatura. **Caso clínico:** Paciente feminina, 30 anos, com diagnóstico de HIV/SIDA e DRC em hemodiálise foi admitida na emergência com queixas de febre, prostração e cefaleia há 6 dias. Ela apresentava um cateter de diálise tunelizado implantado 7 dias antes na veia jugular esquerda. O diagnóstico de SBC foi realizado por tomografia computadorizada abdominal que mostrava um trombo no interior da veia cava inferior o qual estendia-se desde o átrio direito até a veia hepática esquerda. O cateter foi removido e a paciente foi anticoagulada. A paciente estava assintomática no momento da alta hospitalar. **Discussão:** SBC é uma condição clínica rara causada por trombose das veias hepáticas, podendo envolver desde uma até todas as três principais veias. Esta síndrome é em geral associada a desordens mieloproliferativas, a malignidades e a situações de hipercoagulabilidade. Este caso demonstra que o mal posicionamento da ponta do cateter no interior da veia hepática causou esta rara complicação. **Conclusão:** Realização de exame radiológico para avaliar localização da ponta do cateter de diálise é uma medida prudente após cada procedimento, mesmo nos casos de implante de cateter de diálise tunelizados com fluoroscopia.

Palavras-chave: Cateteres venosos centrais; síndrome de Budd-Chiari; trombose venosa.

Introduction

Approximately 80% of patients with chronic kidney disease (CKD) initiate hemodialysis through a central venous cateter.[1,2] The use of central venous catheters vascular access is associated with a high rate of morbidity and mortality.[3-5] Common catheter-related complications reported include high infection rates and central venous stenosis and thrombosis.[3,4]

Vessel wall trauma and endothelial irritation due to repeated cannulation and hyperosmolar solutions and blood flow stasis caused by large catheters in small veins account for the high rate of venous thrombosis. We describe a catheter-related thrombosis leading to Budd-Chiari Syndrome (BCS) due to catheter tip misplacement into inferior vena cava.

Case report

A 30 year-old female patient with history of HIV/AIDS and end-stage CKD on hemodialysis since 2013 was admitted to the emergency department with fever, prostration and headache in the last six days, followed, in the last four days, by unproductive cough, post-prandial abdominal pain and nausea. She had one episode of diarrhea, without pathological features.

In previous medical history, she had five failed arteriovenous fistula and several central venous catheters and catheter-related infections. On admission she had a tunneled dialysis catheter placed at the left jugular vein without phlogistic signs. On physical examination the only abnormal finding was tenderness in right upper quadrant with negative Murphy sign.

Blood tests showed haemoglobin 4.9 mg/dL, leukocytes 10980 (4% immature forms), platelet count 135000/µL, prothrombin time INR of 1.21, partial thromboplastin time 17.8 seconds, creatinine 6.76 mg/dL, urea 64 mg/dL, potassium 4.1 mg/dL, gamma-glutamyl-transferase 164 IU/L, total bilirubin 0.3 mg/dL, alkaline phosphatase 88 IU/L, ALT 31mg/dL, AST 34 mg/dL and C-reactive protein 540 mg/L (reference level < 4 mg/dL), antinuclear antibodies negative, anti-cardiolipin antibody (IgM and IgG) negative.

Chest X-ray was normal with the dialysis catheter tip located in the inferior vena cava (Figure 1). Ultrasound of abdomen evidenced enlarged liver with regular contours and heterogeneous echogenicity, with a predominant high echogenicity surrounded by low echogenicity areas. Hepatic artery shows increased spectral speed (190 cm/s). Abdominal computed tomography (Figure 2) showed a partial thrombus within the inferior vena cava which extended from the right atrium to medium hepatic vein, and continuing along the left hepatic vein. The liver had blunt edges and heterogeneous density, with a predominance of hypodensity with a part of the parenchyma with normal density in the right lobe.

Figure 1. Chest X-Ray showing dialysis catheter tip located in the inferior vena cava (blue arrow).

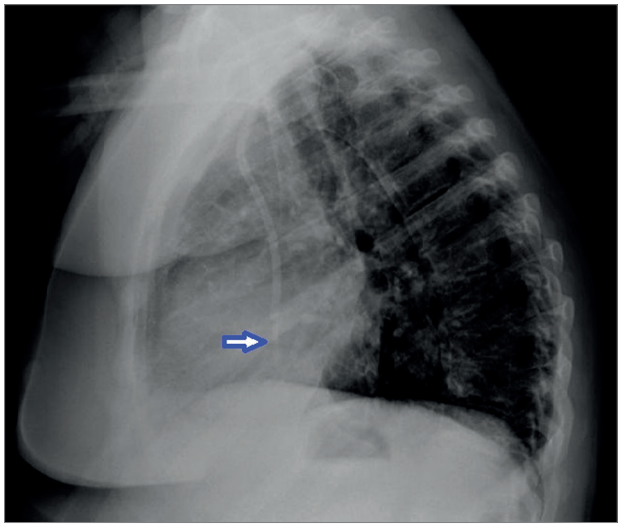
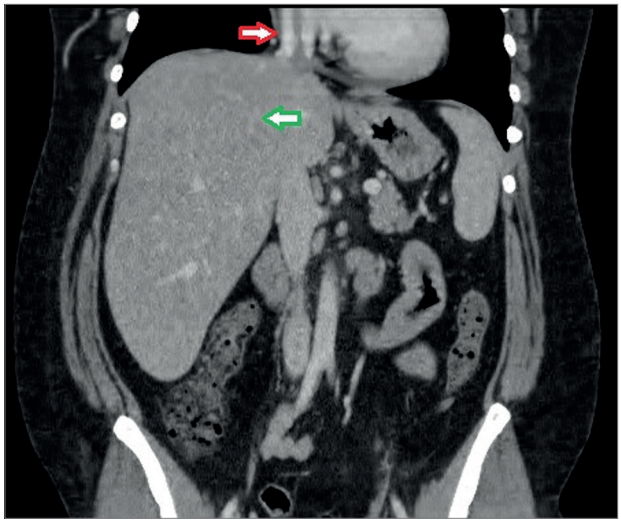


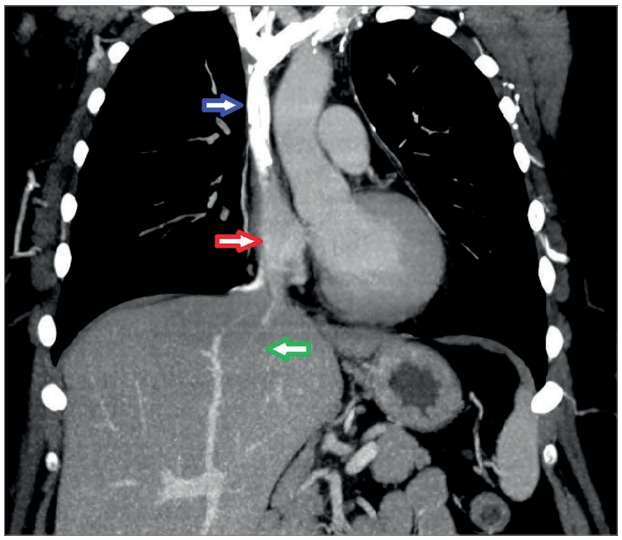
Figure 2. Portal phase of abdominal computed tomography showing subtotal thrombus within the inferior vena cava (red arrow) and a heterogeneous hepatic parenchyma with hypodensity especially at the left lobe (green arrow).



The diagnosis of BCS and catheter-associated infection were confirmed and catheter was removed. Anticoagulation was started with IV heparin followed by warfarin, and infection was treated with vancomycin and cefepime for 14 days.

She was discharged from the hospital assymptomatic. After fifteen months from BCS event, this patient remains in hemodialysis treatment with tunneled dialysis catheter. At this time, abdominal computed tomography showed complete resolution of the thrombus and homogeneous liver parenchyma (Figure 3).

Figure 3. Control abdominal computed tomography fifteen months after the event showing resolution of the thrombus (red arrow), a homogeneous liver parenchyma and a new tunneled catheter (blue arrow) introduced in the left subclavian vein and placed at the superior vena cava.



Discussion

Venous thromboembolism is a common event in CKD patients. These patients present hypercoagulability secondary to several mechanisms such as reduced natural anticoagulant protein levels (i.e. antithrombin), platelet hyperreactivity, and increased blood viscosity.[6,7] Epidemiologic studies have reported an increased risk for VTE in patients receiving dialysis.[8] Both non-tunneled and tunneled hemodialysis catheters are associated with increased risk for venous thrombosis due to higher blood turbulence and endothelial dysfunction - two out of three aspects of Wirchow's triad.

In this case report, patient had been using a tunneled hemodialysis catheter and presented an extensive thrombosis involving inferior vena cava, right atrium, medium hepatic vein, and left hepatic vein leading to BCS. The chest X-ray demonstrated that the tip of dialysis catheter was located in the inferior vena cava. Ideally, central venous catheter tip should lie in the superior vena cava. Although inadvertent introduction of the catheter tip into inferior vena cava is uncommon, it is usually unrelated with major complications.

BCS implies thrombosis of the hepatic veins and/or the intrahepatic or suprahepatic inferior vena cava. Major causes of BCS include myeloproliferative disorders, malignancy and hypercoagulablility states.[9] Few previous studies have described Iatrogenic BCS following supra-hepatic vein cannulation. Fitoz *et al.*[10] described a BCS in a young patient with diagnosis of lymphoma with central venous catheter located into a hepatic vein, and Pieters *et al.*[11] reported a case of transhepatic cannulation of hepatic vein with a catheter for total parenteral nutrition in a child with idiopathic intestinal obstruction.

To our knowledge, it is the first time that BCS caused by misplacement of dialysis catheter tip in a CKD patient on maintenance hemodialysis. Whether other type of thrombophilia contributed to the disease presentation is unknown since complementary tests were not collected, such as factor V Leyden and prothrombin gene mutation. However, presence of a central venous catheter appears to be pro-coagulant enough to trigger such vein thrombosis in this case.

Clinical manifestations of BCS are variable including ascites, hepatomegaly, and abdominal pain. In subacute form, which is the most common presentation, vague abdominal discomfort and unspecific laboratory changes (i.e. discrete transaminases elevation) makes the diagnosis challenging.

BCS should be considered in the differential diagnosis of patients presenting with acute liver failure, acute hepatitis, or chronic liver disease, particularly if the patient has known risk factors for BCS. Diagnosis can usually be established with doppler ultrasonography (US), with a sensitivity and specificity of nearly 85% for BCS.[12]

In this case BCS was not demonstrated by Doppler US, which is an operator-dependent exam, and diagnosis was confirmed with abdominal computed tomography. Symptomatic BCS has a high mortality rate if untreated. Intractable ascites, gastrointestinal bleeding and liver failure are common complications leading to death.

Approach of BCS should consider clinical and anatomic features of each individual patient. In general, treatment includes systemic anticoagulation and management of the complications described above. Thrombolytic therapy and angioplasty can be considered in selected cases.

Dialysis catheters are associated with several complications mainly related to infection and central venous stenosis/thrombosis. This case calls attention for inadvertent catheter tip placement into hepatic vein leading to a severe complication. It is reasonable to check catheter tip location with radiological image after each procedure even if the tunneled dialysis catheter has been introduced with fluoroscopy image.

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