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Case Report

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Secondary Budd-Chiari Syndrome due to Sarcomatous Renal Cell Carcinoma: Case Report

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Abstract

Sarcomatoid renal carcinoma represents 8% of all renal tumors, is caused by any of the renal carcinoma subtypes and is characterized by high cellularity, cellular atypia, and epithelial and mesenchymal components. Sarcomatoid renal cell carcinomas usually have a poor response to systemic treatment, and most patients survive on average 4 to 9 months after diagnosis. Budd Chari syndrome is a rare disease caused by obstruction of the hepatic venous system. If the cause is an endoluminal lesion, it is considered a primary syndrome, while if the cause is external to the venous system, it is considered a secondary syndrome. We present the clinical case of a patient with a sarcomatoid carcinoma that caused a secondary Budd Chiari syndrome with a rapid and aggressive evolution that culminated in multiple organ failure and the death of the patient.

Keywords: Budd-Chiari Syndrome

Abbreviations: BCS: Budd Chiari syndrome; DVT: Deep vein thrombosis; SRCC: Sarcomatous renal cell carcinoma; SAAG: Serum ascites albumin gradient

Introduction

Budd-Chiari syndrome (BCS) is a rare disease caused by the obstruction of the liver venous outflow tract. BCS incidence is of 1-2 cases per 100,000 habitants, the most frequent presentation is in women, in the third and fourth decade of life, and in non-Asian countries the most frequent etiology is the sole obstruction of the hepatic venous system. Hypercoagulable state is diagnosed in 75% of the patients with primary myeloproliferative disease representing the leading cause [1].

BCS is classified according to the site of obstruction, etiology, and clinical presentation. Primary Budd-Chiari syndrome occurs when the obstruction of the hepatic venous tract is due to an endoluminal venous lesion, as thrombosis, endophlebitis or web, while secondary Budd-Chiari syndrome is present when the obstruction is not originated by the venous system, such as malignant tumor invasion, abscess, cysts, or due to extrinsic compression. Budd Chiari syndrome can also be classified based on the site of obstruction, clinical signs, and duration of disease [2].

Case Report

We describe the case of a 52-year-old man with sudden-onset of ascites and abdominal pain with elevated total bilirubin 3.29mg/dL, alanine aminotransferase (227 UI/L), aspartate aminotransferase (729UI/L) and prolonged INR 2.85 (TP 31.9 seg). His medical history was remarkable for an event of deep vein thrombosis (DVT) of the right pelvic limb a year before, which was treated with non-specified oral medications, without having established the etiology of the DVT. Initial diagnostic workup with abdominal ultrasound showed heterogeneous liver echogenicity, decreased portal vein velocity, ascites, and absence of flow into inferior vena cava (IVC); paracentesis showed a serum-ascites albumin gradient (SAAG) of 2.3g, compatible with portal hypertension. Complementary workout included abdominal contrast enhanced CT (CECT) (Figure 1) and 18F-fluorodeoxyglucose (18F-FDG) PET-CT, relevant findings are shown in Figure 2 and Figure 3.





Figure 1: Abdominal CECT. Coronal (A) and axial (B, C) images show a partially defined mass in the right atrium (arrow), acute heterogeneous thrombus in the inferior vena cava (arrow) with extension into the right renal vein (arrow), absence of opacification of the hepatic veins (arrow) with a nutmeg liver appearance, presence of ascites (arrow) and aortocaval node (arrow).



Figure 2: 18F-FDG PET/CT. MIP PET (A) and coronal fused PET/CT (B,C) images show avid 18F-FDG uptake in the right atrium mass, inferior cavo-atrial junction, and aortocaval node (arrows); IVC thrombus showed non 18F-FDG activity.



Figure 3: a) and b) 18F-FDG PET-CT. A) The MIP image shows an increased FDG uptake in the right atrium and right renal hilum. b) In the axial fusion image, the right hiliar uptake is not associated with renal pelvis dilatation (arrow). c), d) and e) Contrast enhanced CT. c) A right atrial tumor (*) is noticed with almost complete lumen obliteration (arrow). d) The coronal reformatted image shows a thrombus who extends into de hepatic veins (*) and the inferior cava (arrow). e) Mild right renal atrophy is noticed (arrow) in association with an ill-defined mass in the renal medulla (arrowhead) and a tumoral thrombus into the renal vein (*).

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Transjugular biopsy of the right atrium mass was performed by the interventional radiology team (Figure 4); histopathology revealed sarcomatoid renal carcinoma emerging from IVC. Absence of desmin, action, CD31 and ERG reactivity discarded leiomyosarcoma and angiosarcoma, an obligation in the presence of this intra jugular growth (Figure 5).







Figure 5: Epithelioid and spindle neoplastic cells admixed with inflammatory infiltrate and edema. The epithelial differentiation was revealed with strong and diffuse reaction with cytokeratin coctail and its kidney origin with the clear nuclear presence of immunoreactivity with PAX-8. All images 400X.

The patient presented KDIGO 3 acute kidney injury with an abrupt decrease in uresis (0.02 ml/kg/hr), hyperkalemia with electrocardiographic changes, and evidence of acute liver failure with hepatic encephalopathy. It was decided to perform renal ultrasound in which an absence of flow in the right renal vein was observed as a consequence of venous thrombosis in the IVC that caused hemodynamic alteration of the renal artery and in the morphology of the ipsilateral kidney, left kidney with decreased vascularity color Doppler application, patent renal vein, hyperdynamic renal artery with increased resistance index. The patient was not a candidate for renal replacement therapy due to the high risk of tumor injury with the placement of the Mahurkar catheter as well as the possible impact of hemodialysis on the patient's hemodynamics by reducing the preload. The sarcomatoid renal carcinoma was unresectable in the background of acute liver and renal failure and the patient subsequently died.

Discussion

There are currently four subtypes of renal carcinoma: clear cell carcinoma, papillary carcinoma, chromophobe carcinoma, and collecting duct carcinoma. Sarcomatoid carcinoma is considered to be a subtype with higher grade undifferentiated transformation which can arise from any subtype of renal carcinoma [3]. There are studies that show that the subtypes that most frequently have sarcomatoid transformation are chromophobic carcinomas, compared to the clear cell carcinoma or papillary carcinoma subtypes; however, due to the greater frequency of clear cell tumors, most cases of sarcomatoid carcinomas seen in clinical practice origin from a clear cell subtype [4]. Sarcomatoid carcinomas are characterized by containing spindle cells, high cellularity, cellular atypia, and epithelial and mesenchymal components; clinically they tend to have an aggressive course and resistance to multiple systemic treatments. Other studies have shown an average survival of 4 to 9 months after diagnosis. They represent approximately 8% of overall renal carcinomas [5].

Whether any subtype of renal carcinoma is at increased risk for developing sarcomatoid transformation is unknown, it is uncertain whether the initial subtype impacts patient prognosis [6]. But it is important to try to subclassify morphologically these lesions, even in diminute samples, due to the potential impact of neoadjuvant treatments. Multiple studies have shown that important prognostic factors in patients with sarcomatoid carcinomas include the presence of lymph node involvement, distant metastasis, and histology-evidenced tumor necrosis [7]. In the case of the patient presented, the sarcomatoid carcinoma caused Budd Chiari syndrome of which we found no reports in the medical literature. There are reports of poor prognosis for patients with this diagnosis, however, none of the reported cases that had a similar evolution to that of the patient we present. At the time of diagnosis, the patient had lymph node involvement as well as distant metastases, which is consistent as a prognostic factor in accordance with what was observed by Ro et al. Despite the fact that new treatments are currently used for patients with clinical stage 4 renal carcinomas, such as sunitinib or sorafenib, these drugs have not improved the prognosis in patients with sarcomatoid carcinoma [8]. The patient presented an excessively rapid progression with evolution to multiple organ failure before being able to obtain the definitive histology result and be able to start palliative management.

Acknowledgement

None.

Conflict of Interest

No Conflict of Interest.

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