



## Neuroendocrine Tumor Mimicking Budd-Chiari Syndrome: A Case Report

Gautam N Mankaney<sup>1</sup>, Parmjeet Randhawa<sup>2</sup>, KV Narayanan Menon<sup>3</sup> and Achuthan Sourianarayanan<sup>4,5\*</sup>

<sup>1</sup>Department of Internal Medicine, University of Pittsburgh Medical Center, USA

<sup>2</sup>Department of Pathology, University of Pittsburgh Medical Center, USA

<sup>3</sup>Department of Gastroenterology, Cleveland Clinic, USA

<sup>4</sup>Department of Gastroenterology, Hepatology and Nutrition, University of Pittsburgh Medical Center, USA

<sup>5</sup>Department of Gastroenterology and hepatology, Medical College of Wisconsin, USA

### Abstract

Diffuse involvement of the liver with neoplastic processes can present a diagnostic dilemma. We present the case of a diffusely infiltrative neuroendocrine tumor (NET) that mimicked Budd-Chiari syndrome (BCS), not previously described in the literature. Histological evaluation can be limited without well-defined margins. Only one of two biopsies revealed a NET. A diagnosis of BCS was first considered after CT and MRI images revealed decreased hepatic and portal venous flow along with liver biopsy suggestive of venous obstruction. However, hepatic venography demonstrated patent veins and another diagnosis was sought. This case illustrates the importance of hepatic venography, the standard imaging technique for diagnosing BCS, and that a NET can mimic BCS.

**Keywords:** Neuroendocrine tumor; Liver mass; Budd-Chiari

### Abbreviations

#### OPEN ACCESS

NET: Neuroendocrine Tumor; BCS: Budd-Chiari Syndrome

#### \*Correspondence:

A Sourianarayanan, Department of Gastroenterology and Hepatology, Medical College of Wisconsin, 9200 West Wisconsin Ave, Milwaukee, WI, 53226, USA, Tel: 414 955 6850; Fax: 414 955 6215;

E-mail: asourianar@mcw.edu

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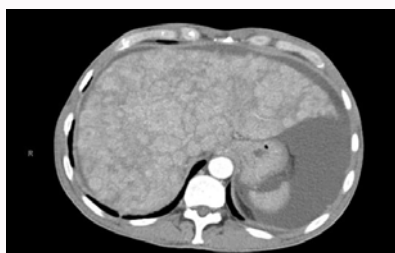
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### Introduction

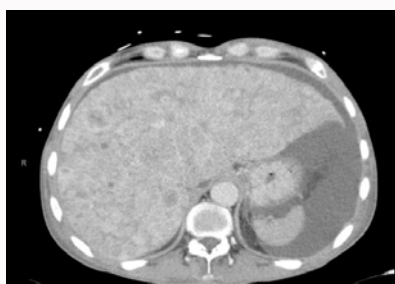
Diffuse involvement of the liver with neoplastic processes can present a diagnostic dilemma. We present the case of a neuroendocrine tumor (NET) of the liver that mimicked Budd-Chiari syndrome (BCS), not previously described in the literature. Diagnosis can be difficult given the rarity and nonspecific presentation of hepatic NETs. Histology and immunohistochemistry remains the most definitive diagnostic modality, which can be limited in diffusely infiltrative NETs by poorly defined margins. In this case, initial biopsy failed to show NET cells and demonstrated venous obstruction as seen in BCS. Follow up hepatic venography revealed patent veins, inconsistent with BCS, and another diagnosis was sought. This case illustrates the importance of hepatic venography, the standard imaging technique for diagnosing BCS, and that a NET can mimic BCS.

### Case Presentation

A 52-year-old male presented with four weeks of abdominal distension and pedal edema. Apart from a 75 pack-years of smoking, his medical history was unremarkable. He had no risk factors for chronic liver disease. Examination revealed tender hepatomegaly, ascites, and pedal edema. His blood chemistry panels were normal. Liver serologies were not elevated but his  $\alpha$ -fetoprotein level was elevated to 17 ng/mL. Doppler ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI) showed compression of the inferior vena cava with absent flow in the middle hepatic and portal veins; MRI also showed flow voids in the hepatic veins. CT revealed numerous liver nodules with some hyperdense enhancement on arterial phase and heterogeneous iso to hypodense enhancement on venous phase, consistent with dysplastic lesions of BCS but also raising concern for hepatocellular carcinoma (HCC) (Figures 1A and B). There were no other masses seen in other organs. On MRI, some nodules were iso to hypointense on T1 weighted images and heterogeneous on T2 weighted images, but few were hyper-intense. Taken together, CT and MRI findings were less consistent with hepatocellular carcinoma but rather regenerative nodules seen in BCS. Ascitic fluid analysis showed no malignant cells and an elevated serum ascites-to-albumin gradient consistent with portal hypertension. Percutaneous liver biopsy revealed diffuse stromal



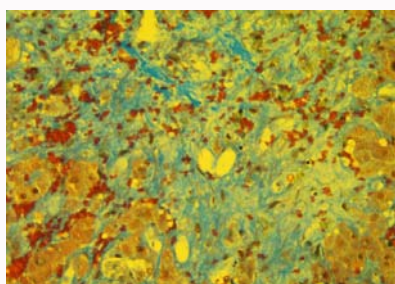
**Figure 1a:** CT image of liver (arterial phase).



**Figure 1b:** CT image of liver (venous phase).



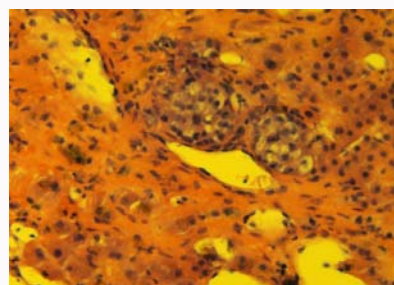
**Figure 2:** The liver biopsy showed centri-zonal hemorrhage, hepatocyte dropout and scattered hemosiderin laden macrophages. H&E stain, x200.



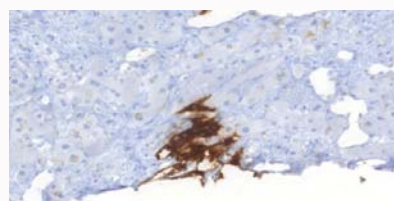
**Figure 3:** The sinusoidal space shows deposition of collagen fibers admixed with extravasated red blood cells. Trichrome stain, x400.

fibrosis and venous congestion with dilated vascular spaces typical of venous obstruction. The patient was diagnosed with BCS. Workup for hypercoagulable conditions was negative, and he was transferred to our institution for further management.

To confirm the diagnosis of BCS, hepatic venography was pursued but revealed patent veins without any evidence of obstruction, inconsistent with BCS. His hepatic venous pressure gradient was elevated to 16 mmHg and repeat biopsy now demonstrated multiple foci of NET cells with background findings suggestive of outflow tract obstruction (Figure 2-4). Immunostains were positive for synaptophysin, chromogranin, and cytokeratin 7 (Figure 5). Serum



**Figure 4:** Two peri-venular clusters of round to polygonal tumor cells with relatively bland nuclei and cytoplasmic vacuoles. H&E stain, x400.



**Figure 5:** The tumor cells show cytoplasmic expression of synaptophysin. Immunoperoxidase stain x400.

chromogranin A level was elevated to 6800 nG/mL and an octreotide scan showed increased isolated uptake throughout the liver; all of these findings were consistent with a NET. There was again isolated liver uptake on positron emission tomography scan.

## Discussion

Primary hepatic NETs are rare, with fewer than 150 reported cases, [1] and differentiating them from other hepatic lesions can be challenging. Our case of a diffusely infiltrative NET mimicking BCS has not been previously.

Our patient's presenting symptoms of abdominal pain, hepatomegaly, and ascites, as well as his IVC narrowing, elevated serum ascites-to-albumin gradient, and initial biopsy were suggestive of hepatic venous outflow obstruction as seen in BCS [2]. Furthermore, radiographic tests demonstrated compression of the IVC with hepatic venous outflow obstruction. Abdominal CT revealed liver nodules inconsistent with hepatocellular carcinoma (HCC) but rather resembles the regenerative nodules associated with BCS. Characteristic CT findings of regenerative nodules in BCS, however, hyper-attenuate on arterial phase and remain hyper-attenuated on portal and venous phases. The nodules are also hyper-intense on T1 and hypo-intense on T2 weighted MRI images [3].

This is a case of NET diffusely infiltrating the liver with obstructed hepatic venules, resulting in venous congesting and mimicking BCS. The absence of tumor on initial biopsy or imaging was also suggestive of BCS. However, hepatic venography demonstrated flow through the hepatic veins, inconsistent with BCS, and concurrent biopsy demonstrated NET cells. This tumor's rarity posed a diagnostic challenge, especially given the variable presentation of NETs in liver and absence of tumor on initial biopsy. One report describes a large primary hepatic NET presenting as BCS by extrinsically compressing the IVC, but its size made it an easy target for biopsy [4]. Diffusely infiltrate NETs are not discrete lesions and can be missed on biopsy, as in our case.

## Author's Contribution

GNM: Manuscript initial design and revision; PR: Pathology slides review, manuscript revision; AS: Design, clinical evaluation, manuscript revision.

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