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Abstract

Background: Over 600,000 people in the United States (U.S.) have liver cirrhosis. It is estimated that 30% of patients with compensated cirrhosis and 60% of patients with decompensated cirrhosis will develop varices in their lifetime, with an incidence rate of new varices at 9% per year. Fifty percent of patients with esophageal varices will experience bleeding at some time. Pheochromocytoma is a rare catecholamine secreting adrenal gland tumor that occurs in 2-8 people per every 1 million people in the general population. Standard treatment for variceal bleeding includes endoscopic band ligation and/or radiologic embolization along with non-selective beta blockers. However, the use of non-selective beta blockers in patients with untreated pheochromocytoma could lead to unopposed alpha receptor stimulation and subsequent risk of life-threatening hemodynamic instability.

Case: A 69-year-old male presented with hematemesis and melena. Emergent upper endoscopy showed bleeding esophageal varices. Band ligation resulted in successful hemostasis. Prior to initiating non-selective beta-blockade (NSBB) for secondary variceal bleeding prophylaxis, he was given doxazosin to minimize unopposed alpha-stimulation.

Conclusion: Variceal bleeding with untreated pheochromocytoma can be safely managed endoscopically and/or radiologically under anesthesia. Providers should administer alpha-blockade in the periprocedural period and prior to initiation of non-selective beta blockers for secondary varices prophylaxis to avoid unopposed alpha receptor stimulation from PCC-related catecholamine secretion.

Keywords

hepatocellular carcinoma, portal vein tumor thrombus, esophageal varices, gastrointestinal bleed, pheochromocytoma, periprocedural management

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Management of Variceal Hemorrhage From Extensive Portal Vein Tumor Thrombus in Hepatocellular Carcinoma and Untreated Pheochromocytoma: A Case Report

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Abstract

Background: Over 600,000 people in the United States (U.S.) have liver cirrhosis. It is estimated that 30% of patients with compensated cirrhosis and 60% of patients with decompensated cirrhosis will develop varices in their lifetime, with an incidence rate of new varices at 9% per year. Fifty percent of patients with esophageal varices will experience bleeding at some time. Pheochromocytoma is a rare catecholamine secreting adrenal gland tumor that occurs in 2-8 people per every 1 million people in the general population. Standard treatment for variceal bleeding includes endoscopic band ligation and/or radiologic embolization along with non-selective beta blockers. However, the use of non-selective beta blockers in patients with untreated pheochromocytoma could lead to unopposed alpha receptor stimulation and subsequent risk of life-threatening hemodynamic instability.

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Keywords

hepatocellular carcinoma, portal vein tumor thrombus, esophageal varices, gastrointestinal bleed, pheochromocytoma, periprocedural management

Abbreviations

CT – computed tomography

GI – gastrointestinal

HCC – hepatocellular carcinoma

MRI – magnetic resonance imaging

NSBB – non-selective beta blocker

PVTT – portal vein tumor thrombus

U.S. – United States

Introduction

Over 600,000 people in the United States (U.S.) have liver cirrhosis.[1] It is estimated that 30% of patients with compensated cirrhosis and 60% of patients with decompensated cirrhosis will develop varices in their lifetime, with an incidence rate of new varices at 9% per year.[2-4] Fifty percent of patients with esophageal varices will experience bleeding at some time.[3,4] Pheochromocytoma is a rare catecholamine secreting adrenal gland tumor that occurs in 2-8 people per every 1 million people in the general population.[5] Standard treatment for variceal bleeding includes endoscopic band ligation and/or radiologic embolization along with non-selective beta blockers.[6] However, the use of non-selective beta blockers in patients with untreated pheochromocytoma could lead to unopposed alpha receptor stimulation and subsequent risk of life-threatening hemodynamic instability.[7] This could also increase the risk of variceal bleeding due to elevated portal venous pressure from alpha-receptor stimulation.[7] We present a rare case that describes the successful management of emergent gastrointestinal (GI) hemorrhage secondary to esophageal varices and severe portal hypertension, in a patient with hepatocellular carcinoma (HCC), portal vein thrombosis extending to the right atrium, and untreated pheochromocytoma.

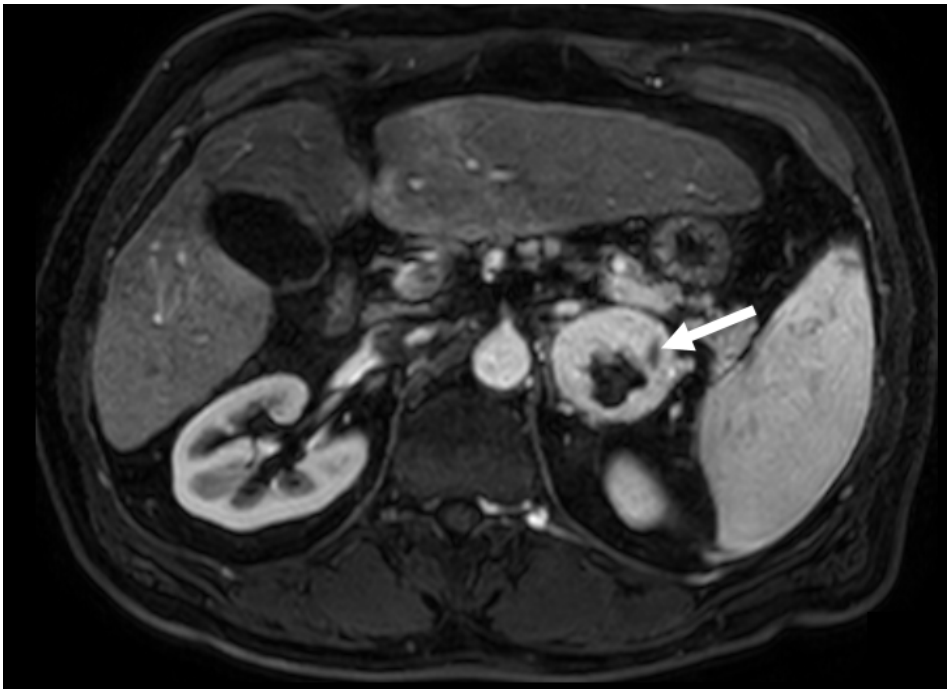

Figure 1. Axial T1 post contrast MRI image, arterial phase: avidly enhancing, heterogeneous left adrenal mass, consistent with a pheochromocytoma (arrow).

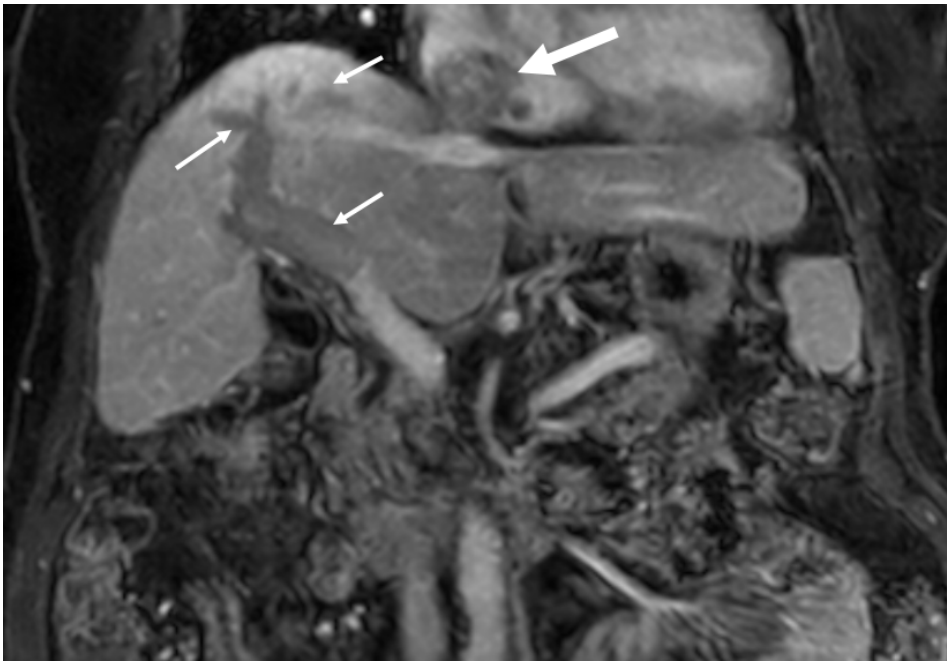

Figure 2. Coronal T1 post contrast MRI image, venous phase: multifocal HCC in the liver dome with a large, enhancing, expansile tumor thrombus in the right and main portal veins (small arrows) and in the right cardiac atrium (large arrow).

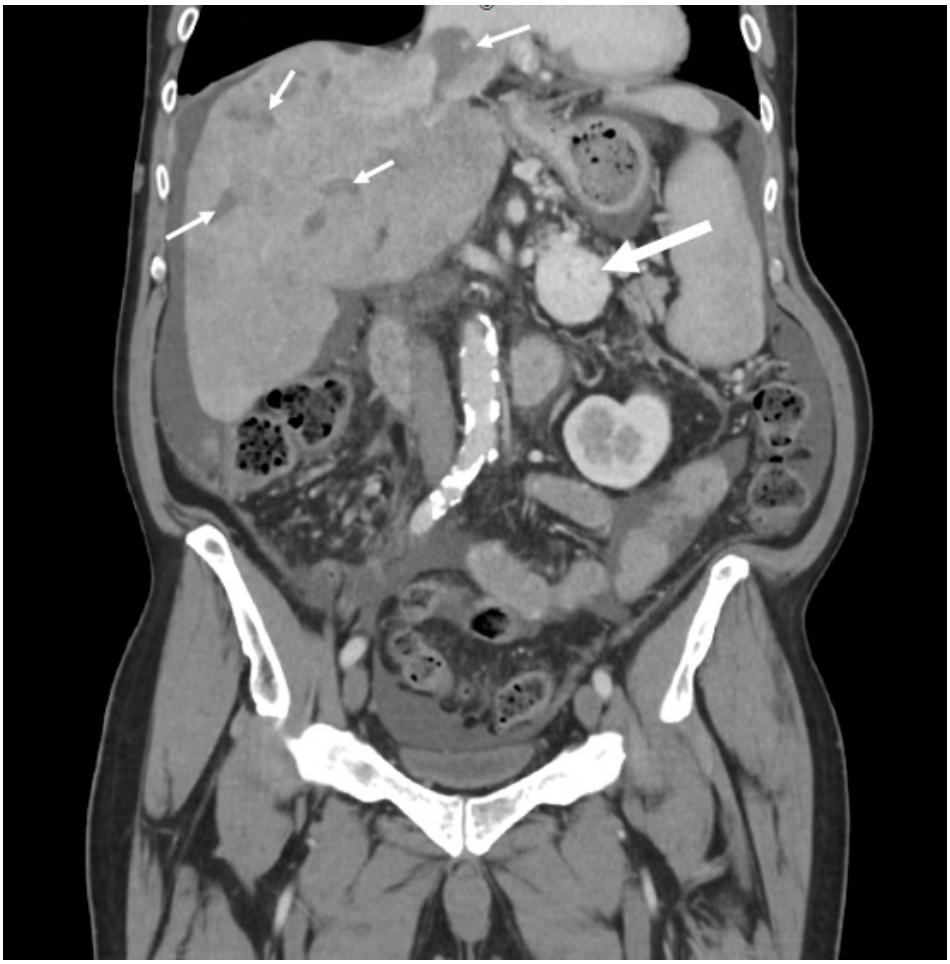

Figure 3. Coronal post contrast CT image, venous phase: multifocal HCC in the liver dome with tumor thrombi within portal and hepatic veins as well as a large tumor thrombus in the right atrium (small arrows). Left adrenal pheochromocytoma is also present (large arrow).

Case

A 69-year-old male presented to the emergency department with acute hematemesis and melena. He has a past medical history of cryptogenic cirrhosis complicated by metastatic HCC, and untreated pheochromocytoma, previously diagnosed by magnetic resonance imaging (MRI) and computed tomography (CT)-guided biopsies. Abdominal imaging following diagnosis revealed tumor thrombus extending from the right hepatic vein through the inferior vena cava entering the right atrium **(Figures 1-3)**. Subsequently, he received systemic chemotherapy but ultimately transitioned to palliative care due to intolerance and disease progression. At presentation, the patient had been untreated for six months.

On physical exam, he was tachycardic (113 beats per minute), hypotensive (blood pressure 92/50 mmHg), tachypneic, and diffusely jaundiced. Laboratory testing noted anemia (hemoglobin 6.4 g/dL down from a prior baseline of 12 g/dL) and total bilirubin 4.6 g/dL. Norepinephrine was initiated for blood pressure support, he was given packed red blood cells, and intubated. He underwent emergent esophagogastroduodenoscopy, which demonstrated four columns of bleeding grade III esophageal varices that were successfully banded with hemostasis achieved **(Figure 4)**. Prior to initiating non-selective beta-blockade (NSBB) for secondary variceal prophylaxis, he was given doxazosin to minimize unopposed alpha-stimulation. However, he experienced episodes of hypotension so doxazosin and NSBB were deferred. Ultimately, he had no further episodes of GI bleeding, was hemodynamically stabilized, and discharged to home hospice.

Discussion

Our cases highlights that variceal bleeding with untreated pheochromocytoma can be safely managed endoscopically and/or radiologically under anesthesia. Providers should consider providing alpha-blockade in the periprocedural period and prior to initiation of non-selective beta blockers for secondary varices prophylaxis to avoid unopposed alpha receptor stimulation from PCC-related catecholamine secretion.

Our patient's acute variceal hemorrhage was complicated for two major reasons. Firstly, periprocedural sedation in pheochromocytoma is complex given the risk of hemodynamic instability and tachyarrhythmias from excessive catecholamine secretion. Although this can typically be mitigated during endoscopy to minimize morbidity and mortality, the

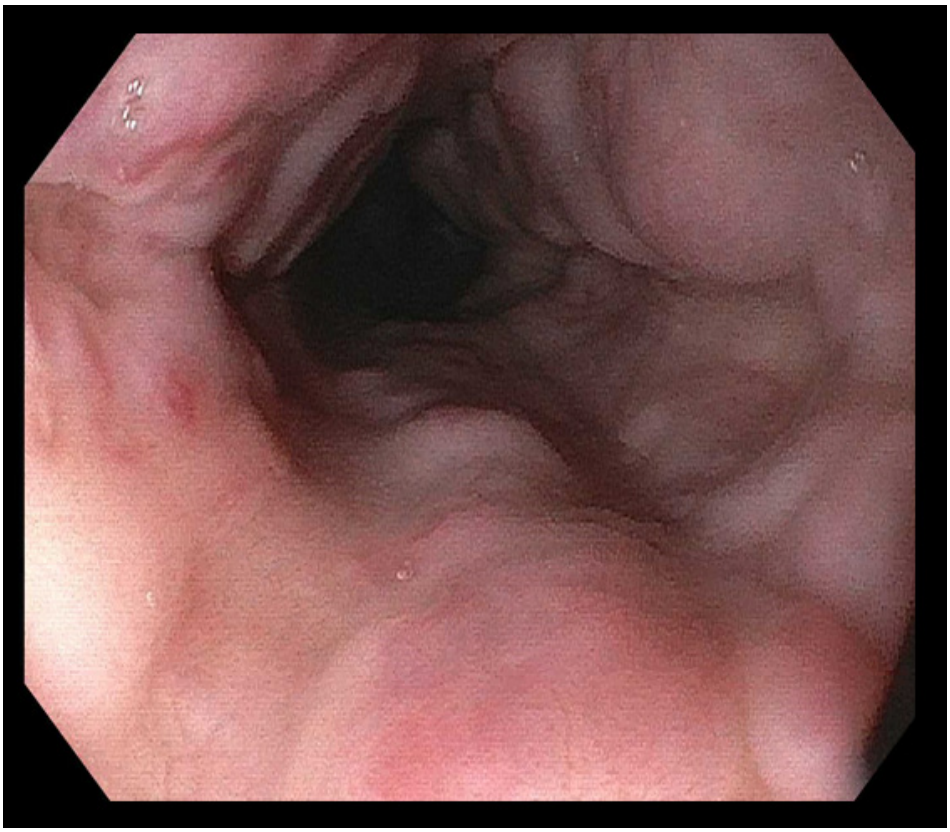

Figure 4. Esophagogastroduodenoscopy demonstrating four columns of grade III esophageal varices in the distal esophagus.

emergent nature of variceal hemorrhage makes timely evaluation and management critical. Secondly, standard of care for variceal bleeding includes endoscopic hemostasis and non-selective beta blockade. However, providing unopposed beta-blockade in the setting of pheochromocytoma can also predispose to hemodynamic instability and tachycarrhthmias.

Our case highlights the complex multidisciplinary care required for successfully managing variceal bleeding in the setting of extensive portal vein tumor thrombus, simultaneous HCC and pheochromocytoma. Successful outcomes in this case were achieved by careful consideration with anesthesia during endoscopy, and with endocrinology for secondary variceal prophylaxis.

Portal vein tumor thrombus (PVTT) is a commonly encountered complication of hepatocellular carcinoma.[8,9] The presence of PVTT affects both treatment options as well as prognosis. The management of PVTT is individualized and depends on comorbidities, bleeding risk, among other factors.[9] The role of anticoagulation in PVTT is unclear. Our case had several complex decision-making points, as he had extensive PVTT through the portal system into the right atrium, putting him at elevated risk for complications of portal hypertension such as hemorrhage, as well as from life-threating thromboembolic events. Given his overall prognosis, personal preference for hospice care, and life-threatening variceal bleeding, the decision was made to withhold anticoagulation.

Given the prevalence of malignancy worldwide and ongoing advances in oncologic care, clinicians will increasingly be exposed to complicated cases such as this and should be aware of optimal treatment strategies, especially in an emergent situation like a GI bleed to maximize successful patient outcomes.

Conclusion

Variceal bleeding in the setting of untreated pheochromocytoma can be safely treated either endoscopically and/or radiologically. Alpha receptor blockade should be provided periprocedurally and before non-selective beta blockers for secondary variceal bleeding prophylaxis to avoid unopposed alpha receptor stimulation from pheochromocytoma. ■

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Conflicts of Interest

All authors report no conflicts of interest. All authors had access to data and had an equal role in writing and revising the manuscript.

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