A Unique Case of Primary Hepatic Neuroendocrine Carcinoma Causing Fulminant Liver Failure.

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Background
* Primary hepatic neuroendocrine tumors (PHNETs) are a rarity and represent about 0.3% of all neuroendocrine tumors.
* We report a rare case of primary hepatic neuroendocrine carcinoma (PHNEC) causing fulminant liver failure in a young male.

Case Report
A 34-year-old healthy African-American male presents with one week of severe RUQ abdominal pain, jaundice, and non-bloody diarrhea. Vitals noted a temp of 98 F, BP 100/60, HR- 108, RR 24 and pulse ox 98% on 2L oxygen. Physical exam revealed yellowing of the skin, scleral icterus, RUQ tenderness, and hepatomegaly. Labs revealed significant leukocytosis with neutrophilia, mild microcytic anemia, sodium 121, chloride 82, bicarbonate 20, total bilirubin (Tb) 11.6, direct bilirubin 7.2, ALP 416, AST 546, ALT 318, albumin 3.1, INR 1.3, LDH 1858, lactic acid 9.8. An abdominal US revealed hepatomegaly with heterogeneous echo pattern, gallbladder wall thickening, CBD dilation at 0.8 cm, ascites, and findings concerning for hepatic vein thrombosis. CT-abdomen with liver mass protocol confirmed above findings.

The patient was then transferred to ICU and started on IV heparin, broad spectrum antibiotics, and vasopressors for hypotension. He had progressive worsening of LFT’s with AST/ALT 4132/1273, Tb 15.5., and INR 1.9. Hepatitis A, B, and C serology were negative, and serum acetaminophen and salicylate levels were normal. N-acetylcystine started for worsening liver failure. Later, he developed AKI with severe metabolic acidosis, and was started on CRRT. Transjugular hepatic venogram and liver biopsy were performed. Venogram demonstrated no filling defects to suggest thrombus. The patient was then transferred to a transplant center for a potential liver transplant. He suffered a fatal cardiopulmonary arrest on the following day. Liver biopsy revealed tumor, immunoreactive with vimentin, and focally with synaptophysin, consistent with poorly differentiated non-small cell carcinoma with neuroendocrine differentiation.

Discussion
* Despite an increase in the incidence of neuroendocrine tumors, PHNEC remains a rarity. Among the reported cases, the highest incidence was in the fifth decade.
* Diagnosis is an evolution, and requires a systematic clinical exclusion with histological confirmation.
* Surgical resection is effective and safe, and prognosis is excellent, despite a high recurrence rate. No clear indication for liver transplantation exists, but can be considered in patients with unresectable lesions and/or fulminant liver failure (1).

Conclusion
* Disease progression is usually slow, but our patient deteriorated rapidly, with multi-organ failure that was fatal.
* PHNECs are very rare tumors, they should be considered as a possible differential diagnosis in the management of hepatic tumors.

References
2. Primary hepatic neuroendocrine tumour: gadoxetic acid (Gd-EOB-DTPA)-enhanced magnetic resonance imaging Acta Radiol Short Rep. 2013 March; 2(2): PMCID: PMC3736966