


# Primary Hepatic Neuroendocrine Carcinoma: Amphicrine Type

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Primary hepatic neuroendocrine carcinoma (PHNEC) is an extremely rare tumor with only about 150 cases reported.<sup>1</sup> PHNECs typically present with vague symptoms such as abdominal pain and occasionally jaundice. This combination of rarity and lack of unique clinical features make the diagnosis of PHNEC difficult. Even more rare are amphicrine carcinomas, defined as those with both glandular and neuroendocrine differentiation in the same cell. Literature review revealed only a handful of case reports of amphicrine carcinomas in the stomach and pancreas and only one other primary to the liver.<sup>2</sup> While there are several treatment options that have been attempted for PHNECs, such as somatostatin analogs, radiotherapy, transcatheter arterial chemoembolization (TACE), and surgical resection; further study of these rare neoplasms is needed to guide definitive treatment and surveillance.

We report a 57-year-old woman whose chief complaint on presentation was abdominal pain. Computed tomography revealed a large mass off segments 5 and 6 of the liver. She underwent extensive workup which included upper and lower endoscopy, as well as a positron emission tomography scan, all of which failed to demonstrate a primary tumor. Subsequent biopsy of the mass was consistent with adenocarcinoma. Due to the location of the mass and lack of primary tumor, the mass was initially presumed to be cholangiocarcinoma. The patient underwent a successful partial right lobectomy and had an uneventful postoperative course. Final pathology was that of a mixed neuroendocrine and non-neuroendocrine neoplasm, amphicrine type, primary in the liver. She was seen at routine follow-up and continues to do well without evidence of recurrence.

Primary hepatic neuroendocrine tumors are rare. Although several treatment options for hepatic neoplasms are available, we propose that surgical resection is the best definitive treatment. A study done by Park et al reported that in 12 patients with PHNEC, those with a single hepatic mass underwent curative surgical resection with free margins had the longest survival rate without recurrence.<sup>1</sup> Those that had multiple lesions or metastatic disease underwent systemic chemotherapy with or without surgical resection. Those who did not undergo surgical resection

showed evidence of recurrence. TACE and other methods of liver protection have been utilized but do not appear to significantly extend the survival time of patients with PHNEC.<sup>3</sup> It is not clear whether the histology of the tumor itself deems the tissue amenable to the effects of certain chemotherapies as there have been so few available for study. For now, however, we propose that surgical resection is the best definitive treatment for these extremely rare carcinomas.

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## References

1. Park CH, Chung JW, Jang SJ, et al. Clinical features and outcomes of primary hepatic neuroendocrine carcinomas. *J Gastroenterol Hepatol*. 2012;27(8):1306-1311. doi:10.1111/j.1440-1746.2012.07117.x
2. Ganesan K, Achmad E, Sirlin CB, et al. Amphicrine carcinoma of the liver. *Ann Diagn Pathol*. 2011;15(5):355-357. doi:10.1016/j.anndiagpath.2010.05.001
3. Zhao Z-M, Wang J, Ugwuowo UC, Wang L, Townsend JP. Primary hepatic neuroendocrine carcinoma: report of two cases and literature review. *BMC Clin Pathol*. 2018;18(3):3 doi:10.1186/s12907-018-0070-7

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