Primary atypical neuroendocrine tumor of the liver: A case report.
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Abstract

Introduction
Primary carcinoid tumor of liver is not a common disease. Primary carcinoid usually reaches a large size at the time of diagnosis.

Case Report
A 58 year-old man submitted to our hospital and abdominal USG revealed a solid mass in liver and a polyp in gall bladder. MRI (Magnetic resonance imaging) showed a 9x8 cm regular shaped hepatic mass at the right lobe posterior segment. At operation hepatic mass was regular shaped and enucleated totally. The neuroendocrine feature was identified by immunohistochemical markers such as synaptophysin and chromogranin.

Discussion
As a result of this case surgery must be in mind as a primer treatment modality. Radiological diagnosis of PHNET can be performed with CT and MRI. There is not a remarkable difference between typical and atypical hepatic carcinoid tumor survival rate.

Introduction
The incidence of primary hepatic neuroendocrine tumor (PHNET) is between 55% and 30% of the digestive system and respiratory system. Commonly PHNETs are seen in the small intestine (45%) in the digestive system. Frequency of PHNET is followed as rectum (20%), appendix (17%), colon (11%) and stomach (7%). Neuroendocrine tumors originated from liver are rare. PHNET usually reaches a large size at the time of diagnosis². Neuroendocrine tumors classified typical and atypical according to the histopathological pattern. Typical carcinoid tumor determined as insular, trabecular, glandular, or mixed pattern. Nuclear atypia, necrotic foci and high mitotic activity seen in atypical carcinoid tumors³. Sometimes it can be difficult to determine whether tumor is primary or metastases. Preoperative radiological studies such as CT and MRI could reveal PHNET. In this study we reported a giant PHNET with radiological and histopathological findings. Because of primary hepatic originate and giant size of tumor this case is original.

Case presentation
A 58 year-old man submitted to our hospital with abdominal distension and pain. He had not history of jaundice and any other digestive symptoms. His family history was unremarkable. Upon physical examination, vital signs were normal. There was hepatomegaly and abdominal tenderness epigastric region. Biochemical tests were normal. Tumor markers including α-Fetoprotein (AFP), carcinoembryonic antigen (CEA) and CA 19-9 were within normal limits. Blood hepatitis B and hepatitis C tests were negative. Abdominal USG revealed a solid mass in the liver and a polyp in gallbladder. MRI (Magnetic resonance imaging) showed a 9x8 cm regular shaped hepatic mass at the right lobe posterior segment (Figure 1). Based on radiological findings, surgical team decided to perform surgery. At operation 10x10 cm measured hepatic mass was regular shaped and enucleated totally. There was no evidence of tumor at the rest of the liver and other organs. Ten days after operation patient discharged from hospital. The tumor was composed of a monomorphic cell population arranged in a predominately solid with some acinar (Figure 2A) and trabecular architecture. The stroma was hyalinized in some areas. Cytologically, the cells showed minimal atypia and possessed eccentric, speckled nuclei with cytoplasmic granularity and eosinophilia. The tumor cells also exhibited atypical features in the form of increased mitotic activity and nuclear hyperchromasia. The neuroendocrine feature was identified by immunohistochemical markers such as synaptophysin (Figure 2B) and chromogranin.

Discussion
Atypical primary neuroendocrine tumor of the liver is not a common clinical practice. Female cases are four times more than male. Usually patients are present with upper abdominal distention and pain. At the time of admission...
the hospital hepatomegaly develops nearly in all of the patients. In our case he had hepatomegaly and abdominal distention in initial examination. Only in few cases (10%) reported that patients were asymptomatic. Most of asymptomatic cases were smaller sized less than 3 cm. Hepatic neuroendocrine tumor could be sized up to 20 cm\(^4\). Usually primary diagnostic method for liver masses is abdominal ultrasound. When ultrasound revealed a mass in the liver, computed tomography (CT) or magnetic resonance imaging (MRI) performed as advanced radiological procedures to determine the originate of mass\(^5\). CT and MRI can help surgical teams for surgical strategy. In this case we performed an MRI. The most frequent therapy of neuroendocrine tumor is surgical resection. Surgery is performed 85% of primary hepatic neuroendocrine tumor\(^6\). We decided to perform total excision of hepatic mass by reason of regular shaped and well-defined appearance. Hepatic chemoembolization and radiofrequency ablation can be a useful therapy for unresectable tumors that is limited in the liver. Five-year survival rate of primary hepatic carcinoids is 92%; survival rate is decreased to 45% with the existence of metastasis\(^7\). There is not a remarkable difference between typical and atypical hepatic carcinoid tumor survival rate.

**Conclusion**

Here we reported a primary atypical hepatic carcinoid with a giant size. As a result of this case surgical treatment must bear in mind as a primer treatment modality. Radiological diagnosis of PHNET can be performed with CT and MRI.

**References**


![Figure 2A](image2.png)

**Figure 2A:** The tumor cells were monomorphic and arranged in a predominately solid with some acinar architecture (H+E X20).

![Figure 3B](image3.png)

**Figure 3B:** There was positivity with synaptophysin (H+E X40).

![Figure 4B](image4.png)

**Figure 4B:** The tumor cells were monomorphic and arranged in a predominately solid with some acinar architecture (H+E X20).