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Primary Liver Neuroendocrine Tumor : A Case Report And Literature Review

Sanghwa SONG[1], Yangseok KOH*[1], Cholkyoon CHO[1], Heejoon KIM[1], Eunkyu PARK[1], Younghoe HUR[1]

[1]Division Of Hepato-Pancreato-Biliary Surgery, Department Of Surgery, Chonnam National University Medical School, REPUBLIC OF KOREA

Background : Neuroendocrine tumors (NETs) are low-grade malignancies arising from neuroendocrine cells. Primary hepatic neuroendocrine tumors (PHNETs) are extremely rare and difficult to differentiate PHNETs from other liver tumors, such as hepatocellular carcinoma (HCC) or cholangiocarcinoma.

Methods : A 22-year-old man presented with a complaint of intermittent abdominal pain lasting. Blood tests showed normal values including alphafetoprotein (AFP) and PIVKA-2 as tumor markers and negative hepatitis B and C serology. On ultrasonography (US), a 5 × 4 cm mixed echogenic mass was observed, which was considered as hemangioma or other liver tumor. On abdominal computed tomography (CT), the arterial phase showed a 5.1 cm heterogeneously enhancing mass in the S6 of the liver with a delayed washout pattern in the portal phase. Subsequently performed magnetic resonance imaging (MRI) revealed a 5.0 cm well-defined solid mass and a cystic mass lesion, and the solid mass showed arterial hyperenhancement and a delayed washout pattern in the portal phase; these MRI findings strongly indicated HCC. Based on a preoperative diagnosis of HCC of the right liver lobe, laparoscopic right hepatectomy was performed.

Results : Histopathologically, a 6.6 × 4.2 cm yellow to brown solid mass was found in S6, and no tumor cells were present at the surgical margins. Immunohistochemically, tumor cells were positive for epithelial markers including cytokeratin as well as neuroendocrine markers including CD56, synaptophysin, chromogranin A (CgA), and neuron-specific enolase, with a Ki-67 labeling index of 1–2%. A final diagnosis of carcinoid tumor, i.e. grade 1 NET, was rendered.

Conclusions : PHNET is an extremely rare tumor, which is difficult to diagnose by imaging alone due to its similarity to other liver tumors. Since the differentiation between metastatic NET and PHNET is demanding by histopathology alone, it is necessary to confirm the absence of additional lesions in the gastrointestinal tract by endoscopy, and octreotide scan and serum CgA level can be useful for differential diagnosis. Various techniques such as surgical resection, TACE, and RFA can be applied for the treatment of PHNET. The prognosis of patients with PHNET after treatment can vary according to tumor grade as seen in those with NET; grade 1 PHNET has good prognosis after hepatectomy, whereas grade 3 NETs include unresectable cases and have a poorer prognosis than grade 1 tumors.

Corresponding Author : **Yangseok KOH** (yskoh@chonnam.ac.kr)