



A Case of Primary and Multiple Hepatic Neuroendocrine Carcinomas

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Abstract

Primary Hepatic Neuro Endocrine Carcinomas (PHNECs) are extremely rare. The clinical features and treatment outcomes are not well understood due to rarity of the tumors. We reported on a case of primary and multiple hepatic neuroendocrine carcinomas. The course of disease, diagnostic modality, treatment, surveillance and prognosis were discussed with literatures.

Case Presentation

A 65 year-old female with history of left breast cancer (infiltrating ductal carcinoma), pT2N1M0 underwent modified radical mastectomy 14 years ago. She received adjuvant chemotherapy with regimen of CEF (cyclophosphamide, epirubicin, 5-FU) for six courses and subsequent hormone therapy (tamoxifen for 5 years). She has no history of hepatitis or wine consumption but ever received cholecystectomy and choledocholithotomy for symptomatic biliary stones 12 years ago. However, she was noted to have multiple hepatic lesions while follow-up routinely two years ago (Figure 1 and 2). She got diarrhea and hot flush intermittently in the period. We performed core biopsy for liver tumors and the pathology reported as neuroendocrine carcinoma (Figure 3). The serum investigation revealed high level of chromogranin-A and gastrin. She received gastroscopy without obvious ulcer noted. The patient received partial hepatectomy and radiofrequency ablation for liver tumors (Figure 4). At laparotomy, we did not find any other lesion in so-called gastrinoma triangle. We prescribed long-acting release octreotide (30mg, intramuscular injection, monthly) as adjuvant treatment for one year. Multiple metastases to bone and hilum of right lung occurred to her eight months after surgery (Figure 5). Currently, she is in stable disease with cytotoxic chemotherapy (cisplatin and 5-FU) and surveillance for serum chromogranin-A and gastrin.

Discussion

Primary Neuroendocrine Tumors (NETs) of the liver are extremely rare [1-4]. According to 2010

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Figure 1: The liver MRI revealed one hypersignal nodule in the liver, S5.



Figure 2: The liver MRI revealed two hypersignal nodules in the liver, S4, 7.

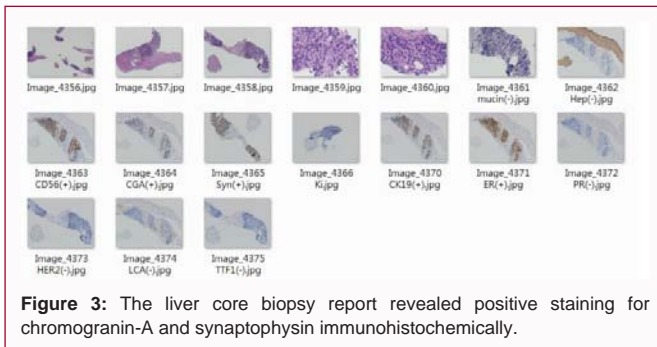


Figure 3: The liver core biopsy report revealed positive staining for chromogranin-A and synaptophysin immunohistochemically.

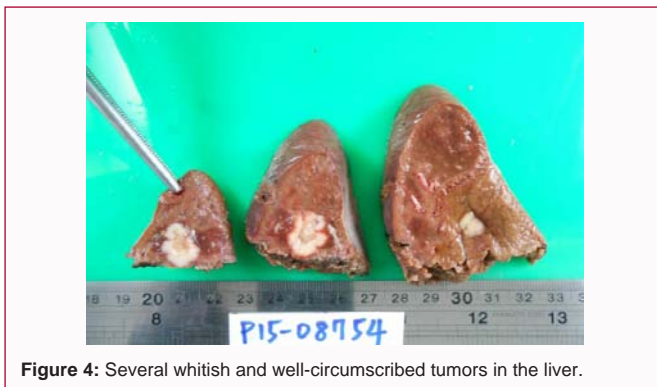


Figure 4: Several whitish and well-circumscribed tumors in the liver.

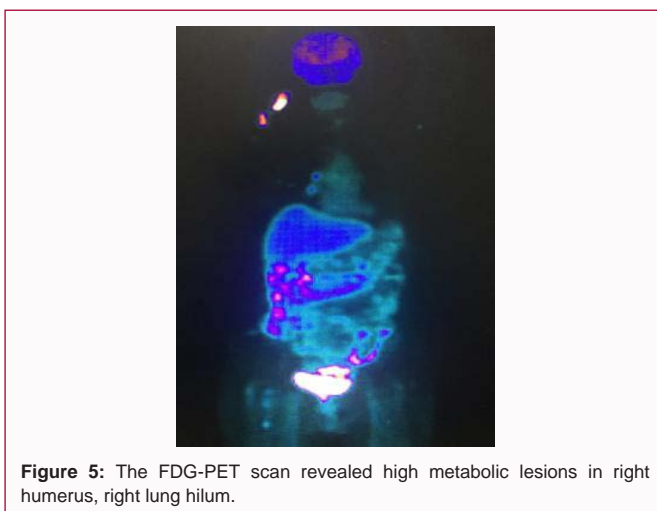


Figure 5: The FDG-PET scan revealed high metabolic lesions in right humerus, right lung hilum.

World Health Organization classification, NETs were categorized according to the clinicopathologic features, treatment condition, and prognostic factors [1]. There was a marked male predominance either in gastroenteropancreatic NETs with liver metastasis or primary hepatic NETs [1]. Primary hepatic NET is mostly single

lesion while the metastatic hepatic NET is mostly from pancreas with multiple nodules in both lobes, with aggressive malignancy and poor prognosis [1]. The prognosis is different in primary and metastatic NETs with 5-year Disease-Free Survival (DFS) rates of 30% and 40% and 5-year survival rates of 35% and 66%, respectively [1]. Surgery is an effective method for the treatment of hepatic NETs and tumor grading is an important determinant factor of prognosis [1,3]. Alternative treatment is transcatheter arterial chemoembolization (TACE) [2,4-6]. There is a reported case in Korea for a 41 year-old woman with primary neuroendocrine carcinoma (NEC) and multiple liver metastases [2]. That patient was misdiagnosed as multiple liver hemangiomas previously and received TACE using a combination of oxaliplatin, gelatin sponge particles and lipiodol [2]. Similarly, our presented case is a primary hepatic NEC with liver metastases without other notable origination. We treated her with cytoreductive surgery including palliative hepatectomy, RFA. We prescribed target therapy for symptomatic relief and chemotherapy for disease control. Primary hepatic neuroendocrine carcinoma should be considered as a possible differential diagnosis for hepatic tumors. The liver can be the primary origin of neuroendocrine tumors, and surgical resection must be considered for curative treatment for primary lesions. However, the prognosis is dismal while recurrence or distal metastasis.

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