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A case report

Primary Hepatic Carcinoid tumor: Dynamic CT and MRI findings

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الورم الكارسنويدي الاولي في الكبد: تقرير حالة

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أستاذ مساعد جامعة أم القرى, كلية الطب, استشاري أشعة الجهاز الهضمي والبولي والتناسلي واستشاري ألاشعة الجراحية.

الملخص العربي

الورم الكارسنويدي الاولي في الكبد يعتبر من الاورام النادرة جدا وعدد قليل منها قد تم توثيقة ونشرة في المجالات العلمية بعدد لا يتجاوز 60 حالة فقط.
نوجز هنا حالة مسجلة لورم كارسنويدي اولي في الكبد لمريض يبلغ من العمر 42 سنة قدم بأعراض الألم في البطن والقى والاسهال. تم تشخيص حالتة بأنة ورم سرطاني كرسنويدي أولي في الكبد علي اساس التحاليل الاشعاعية والمختبرية. تم إكتشاف ورم كبير في الفص الايسر من الكبد (الجزء 2 و3) ومستويات مرتفعة بشكل ملحوظ من (5-HIAA) في البول. أثبتت تحاليل الانسجة بعد الاستئصال الورم السرطاني الكارسنويدي الاولي في الكبد.

ABSTRACT

A 42-year-old man presented with abdominal pain, vomiting, and diarrhea. He was diagnosed with primary hepatic carcinoid tumor based on radiologic, laboratory and histopathological findings. Investigations showed a large mass in the left lobe of the liver (segment 2 & 3) and markedly elevated levels of 5-hydroxyindole acetic acid (5-HIAA) in the urine. Histologic and immunohistochemical findings of the resected left liver lobe mass showed a malignant carcinoid tumor. This case is of interest because of the rarity of this neoplasm. This case report describes and reviews the radiologic dynamic Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) features of this rare neoplasm.

Keywords: *Primary hepatic carcinoid tumor (PHCT), Focal Nodular Hyperplasia (FNH), Dynamic computed tomography, Dynamic Magnetic Resonance.*

INTRODUCTION

Carcinoid tumors develop from neuroendocrine cells and occur most frequently in the gastrointestinal tract in about 85% of cases, the respiratory system in 10% of cases and the rest inother various organs [1]. The vast majority of carcinoid liver lesions are metastatic in nature, but primary hepatic carcinoid tumors are extremely rare. Only few cases were reported since its first description by Edmondson in 1958 [2].

METHODS

A 42-year-old man presented to the surgery outpatient clinic with history of vomiting and diarrhea. The history also revealed abdominal pain over the last 6 month which was becoming more severe in the last two months before the presentation.

Physical examination demonstrated possible abdominal midline mass; otherwise the physical examination was normal with no clinical features of cirrhosis. Blood testing demonstrated mild anemia, however, the results of other laboratory investigations, including liver function tests and blood levels of tumor markers including α-fetoprotein, Ca-19-9, and carcinoembryonic anti-gen (CEA) were within normal limits.

Urinary excretion of the 5-HIAA over 24 hours sample collection (70mg/24h) was markedly elevated. The patient was referred to the radiology department for further evaluation of the midline abdominal mass by CT examination. The Non-contrast CT examination demonstrated a well encapsulated large left hepatic lobe lesion, involving segment 2 and 3 of the liver and measuring 12x13cm with central areas of low attenuations. (Fig.1A).

Subsequently, the patient went for a dynamic CT scan of the liver as per standard protocol. Arterial, Porto-venous and delayed images were obtained, and multiplanar reconstruction was performed.

On the arterial phase, avid enhancement of the lesion was seen with central frond-like projections were seen of low density (Fig. 1B). Direct supply of the lesion was seen by the left hepatic artery which was engorged and larger than the right hepatic artery (Fig 2A & B). On the porto-venous phase of the liver, the lesion showed no significant washout, and the central areas of low attenuations were found more conceptious and of lower attenuation relative to the rest of the lesion (Fig. 1C). Furthermore, the lesion showed well seen porto-venous supply, which was driven via the left portal vein, that wraps around the lesion and sends its branches into the lesion (Fig. 2C & D). On the delayed images, the lesion becomes homogeneous to the liver parenchyma with no change in the central frond like projections of low attenuation. (Fig. 1D).

At this stage and so far, the lesion was not behaving of a classical benign lesion and the central area of low attenuation further raised the suspicion as necrosis would be the main concern. This particular feature was more suggestive of either a primary malignant liver lesion or a metastatic lesion. After discussion with the referring surgeon, further evaluation by MRI was carried out especially, given the fact that Fibro Nodular Hyperplasia (FNH) was still in the differential diagnosis, although uncommon for the patient's gender.

On T1 weighted images, the lesion was encapsulated and shows inhomogeneous low signal intensity with lower signal intensity is seen centrally (Fig. 3A). On T2 weighted images, the entire lesion was of high signal intensity but centrally the lesion was of higher signal relative to the rest of the lesion (Fig. 3B). On the dynamic MRI post Gadolinium administration, the central area remained of low signal intensity throughout theglucagon like peptide dynamic imaging

with more homogeneous enhancement seen at the periphery of the lesion with a capsular enhancement seen persistently in all phases of the dynamic study with similar enhancement pattern to the dynamic CT examination seen on the arterial and the porto-venous phases (Fig. 3 C & D). The CT and MRI showed no radiologic features of liver cirrhosis. The patient refused the biopsy to obtain a tissue diagnosis as he was worried about the spread of the tumor. Given the fact that the CT and the MRI results were not reassuring and given the fact that the patient 5-HIAA was markedly elevated, the patient was booked for left hepatectomy. Meanwhile, more thorough investigations to rule out the possibility of metastatic carcinoid were performed; including upper and lower gastrointestinal endoscopies, CT Enteroclysis, CT chest, and Octreotide scintigraphy. They were all negative except for a focal uptake seen in the liver lesion on the Octreotide uptake scan. Surgery was performed, and the resected left lobe contained a solid tumor measuring 12x13cm (Fig. 4A, B, C & D). The microscopic and immunohistochemical findings were consistent with malignant carcinoid tumor of the liver. The final diagnosis in this case was primary hepatic carcinoid tumor. Annual follow-up of the patient over the last six years showed no local recurrence or metastatic disease.

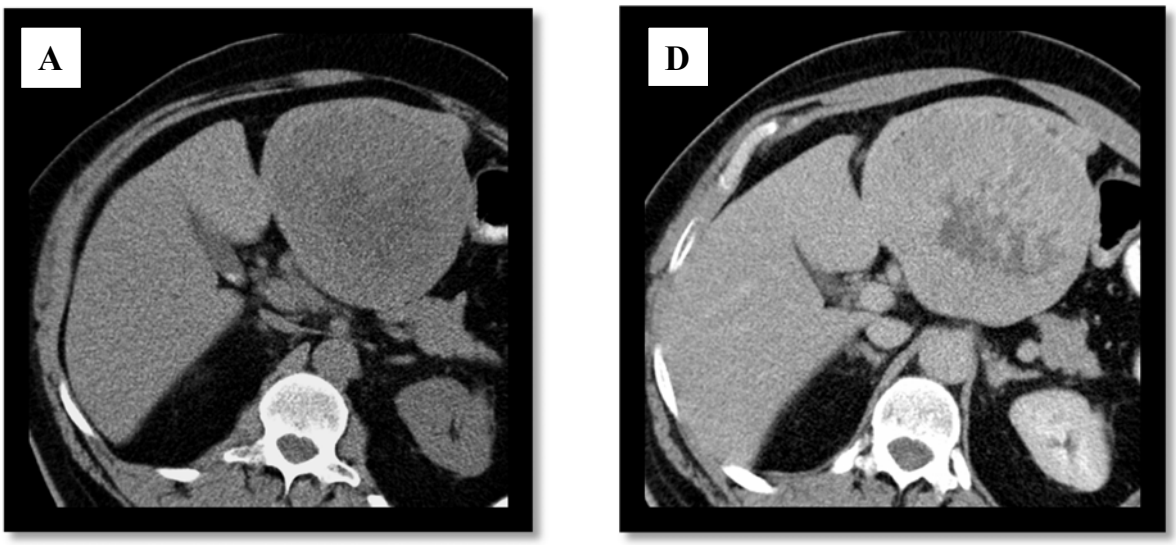

Figure 1. Dynamic Computed Tomography of the liver

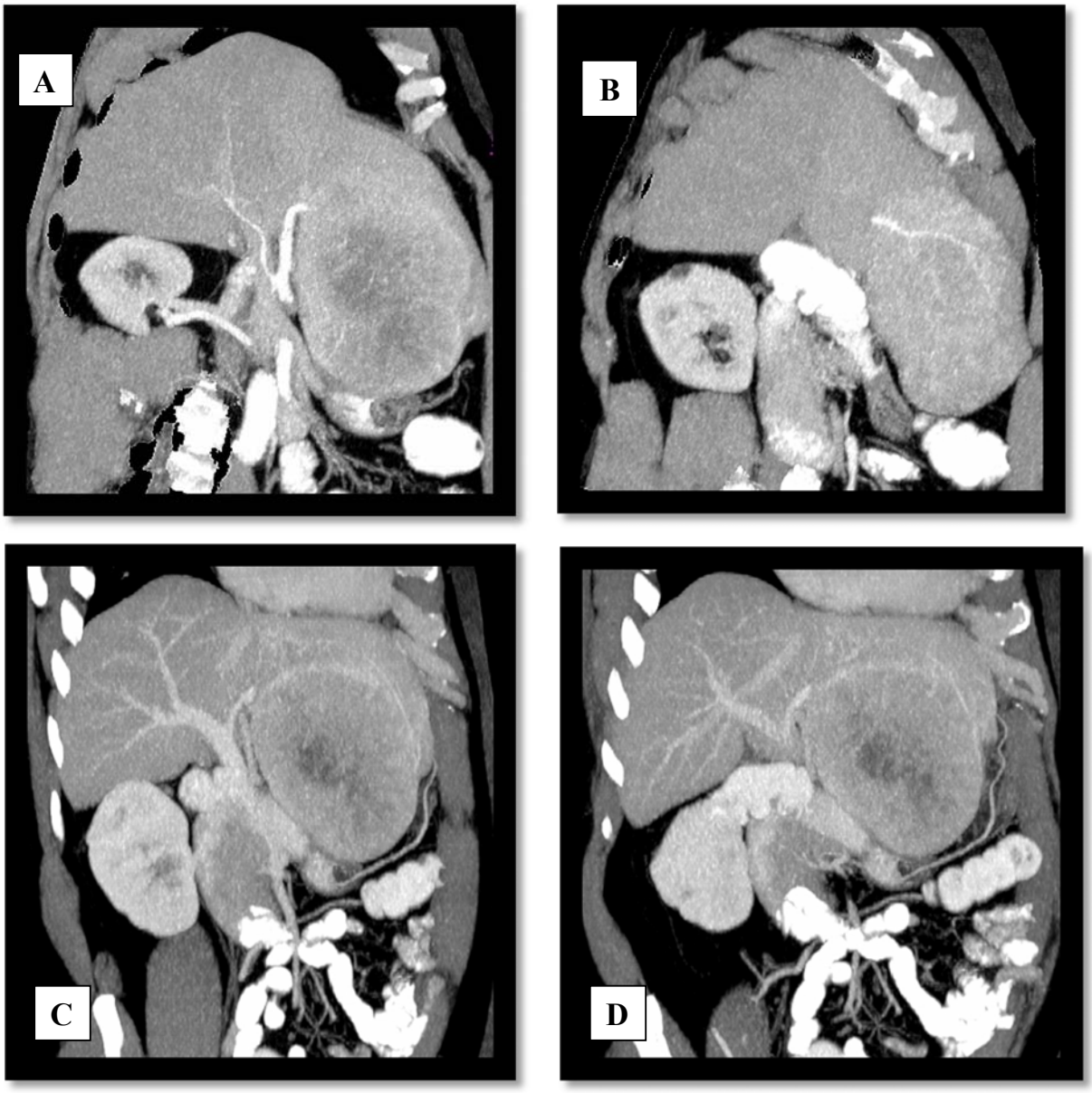

Figure 2. Reconstructed Dynamic Computed Tomography of the liver

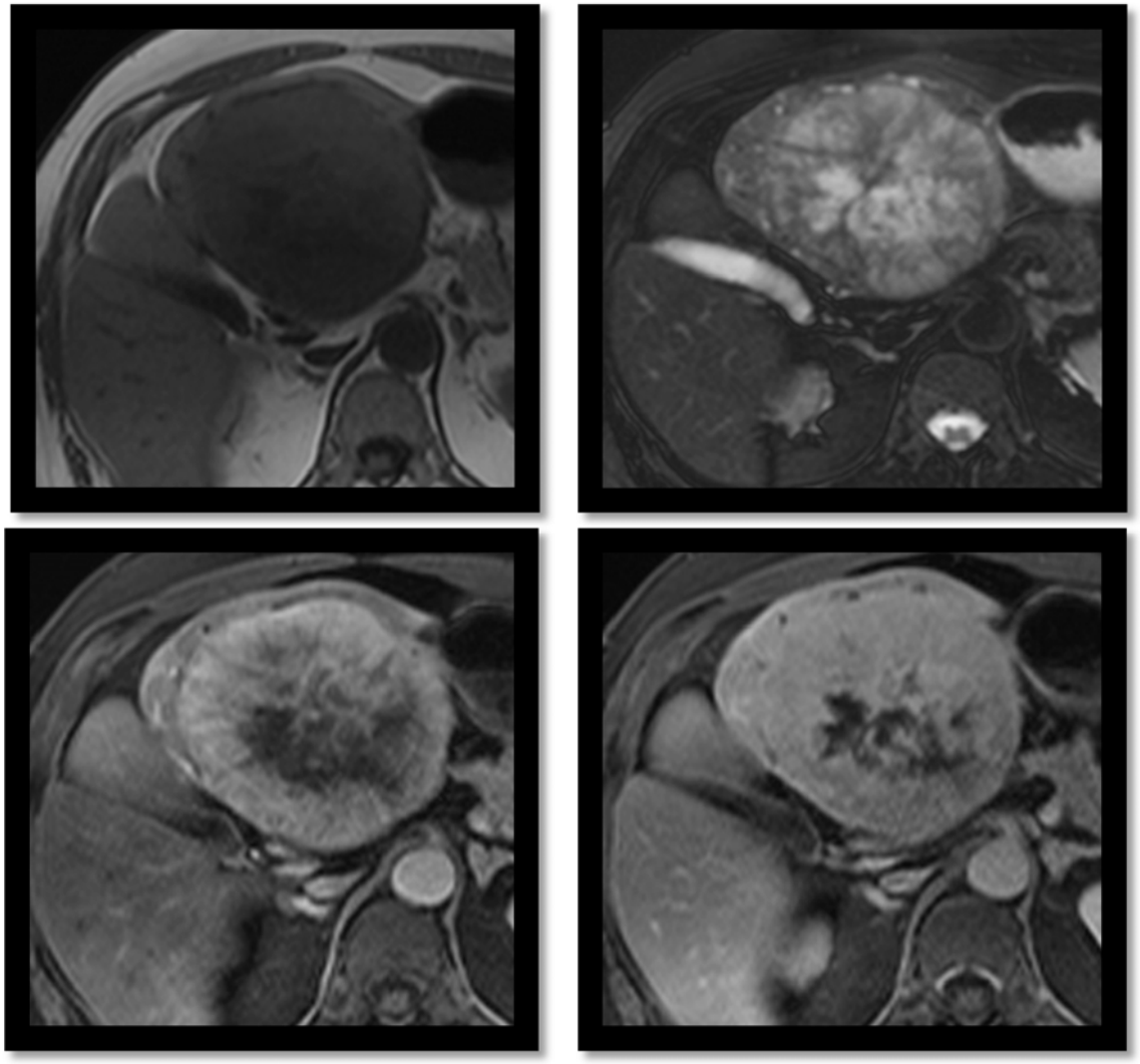

Figure 3. Dynamic Magnetic Resonance of the liver

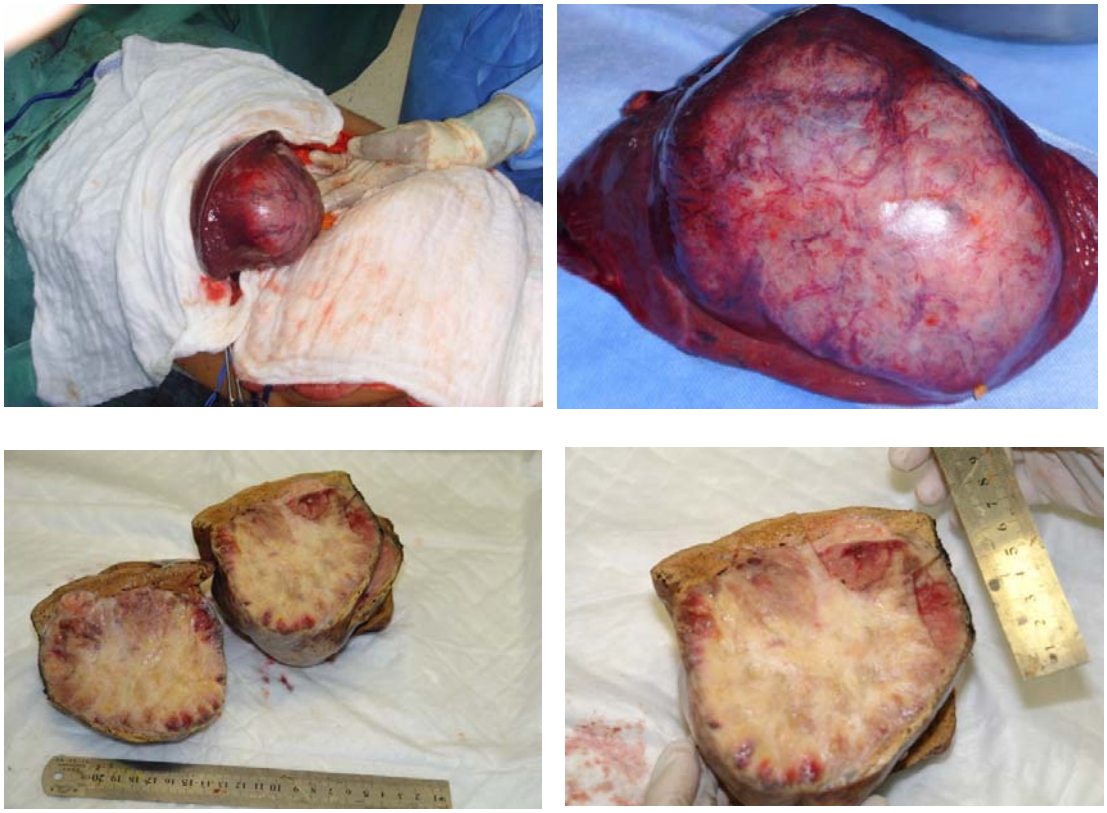

Figure 4. Gross specimen of the PHCT

DISCUSSION

Carcinoid tumors are rare slowly growing neuroendocrine neoplasms. Liver is the most common site for neuroendocrine tumor metastases, but primary hepatic carcinoid tumors are extremely rare [3]. There are about 60 cases reported in the literature [14]. On the previous reports, the CT and MRI findings concur with the findings of this report, but the features of these lesions are atypical and still can be confused with other malignant lesions such as Hepatocellular Carcinoma or benign lesions such as Focal Nodular Hyperplasia [4, 5, 6, 7 8, 9]. The main benign differential diagnosis in the present case was fibro nodular hyperplasia, which is a benign vascular hepatic neoplasm that is most prevalent in young women. The lesions consist of hepatocytes, bile ducts, blood vessels, and Kupffer cells and are characterized by scar tissue in the center [10]. Central area of low attenuation is seen in almost all of the previously reported primary liver carcinoid tumors, but their etiology differs as they were found to be related to necrosis within the lesion or related to central scaring [11]. In this reported case, the central area of low attenuation was the result of necrosis within the lesion, and this was clearly demonstrated on the MRI and pathologically correlated. The presence of a capsule was described and also identified in this case report. [4]. The majority of patients on the earlier reports were hormonally inactive [5], but in this report, the patient suffered from features of carcinoid syndrome, which were further

confirmed by the laboratory results that normalized after resection of the tumor.

Hormonally active tumor releases vasoactive substances into the systemic circulation. Serotonin (5-Hydroxtryptamine) is considered the main vasoactive substance responsible for the majority of symptoms encountered in patients with carcinoid syndrome; such as diarrhea, wheezing and those related to fibrotic reactions in the heart [12, 13]. Elevated metabolic breakdown of this active substance in the urine, namely the 5-Hydroxyindoleacetic acid over 24h is considered a highly reliable method for diagnosis [12, 13]. We noted that the majority of the previously reported primary hepatic carcinoid tumors were found in the right lobe of the liver, contrary to this case in which the lesion was in the left lobe of the liver.

CONCLUSION

Although common findings were observed in the literature for the reported cases of primary hepatic carcinoid tumors and agree with this report, the features are not specific and can be seen in benign and malignant lesions of the liver. Therefore, atypical lesions should be further evaluated by biopsy, and thorough investigations are strongly recommended if the carcinoid tumor is revealed. This is particularly true given the fact that the majority of carcinoid lesions in the liver are related to metastatic deposits. Treating the liver lesion without addressing the primary will have dire consequences.

REFERENCES

1- Kehagias D, Moulopoulos L, Smirniotis V, et al. (1999). Imaging findings in primary carcinoid tumour of the liver with gastrin production. Br J Radiol 72:207–209

2- Iimuro Y, Deguchi Y, Ueda Y, et al. (2002) Primary hepatic carcinoid tumor with metachronous lymph node metastasis after longterm follow up. J Gastroenterol Hepatol 17:1119–1124

3- Furrer J, Hattenschwiler A, Komminoth P, et al. (2001) Carcinoid syndrome, acromegaly, and hypoglycemia due to an insulin secreting neuroendocrine tumor of the liver. J Clin Endocrinol Metab 86:2227–2230

4- M. van der Hoef, D. W. Crook, B. Marincek, D. Weishaupt, et al. (2004) Primary neuroendocrine tumors of the liver: MRI features in two cases. Abdom Imaging 29:77–81.

5- Pilichowska M, Kimura N, Ouchi A, et al. (1999) Primary hepatic carcinoid and neuroendocrine carcinoma: clinicopathological and immunohistochemical study of five cases. Pathol Int 49:318–324.

6- Ruckert RI, Ruckert JC, Dorffel Y, et al. (1999) Primary hepatic neuroendocrine tumor: successful hepatectomy in two cases and review of the literature. Digestion 60:110–116.

7- Kehagias D, Moulopoulos L, Smirniotis V, et al. (1999) Imaging findings in primary carcinoid tumour of the liver with gastrin production. Br J Radiol 72:207–209.

8- Tjon ATRT, Jansen JB, Falke TH, Lamers CB. (1994) Imaging features findings in primary carcinoid tumour of the liver with gastrin production. Br J Radiol 72:207–209.

9- Sofka CM, Semelka RC, Marcos HB, Woosley JT. (1997). MR imaging of

metastatic pancreatic VIPoma. Magn Reson Imaging 15:1205–1208.

10- K. J. Mortelé, M. Praet, H. Van Vlierberghe, M. Kunnen, P. R. Ros. (2000) CT and MR Imaging Findings in Focal Nodular Hyperplasia of the Liver Radiologic—Pathologic Correlation. AJR vol. 175 no. 3 687-692.

11- Iwao M, Nakamuta M, Enjoji M, et al. (2001) Primary hepatic carcinoid tumor: case report and review of 53 cases. Med Sci Monit 7:746–750.

12- Hsueh C, Tan XD, Gonzalez-Crussi F. (1993) Primary hepatic neuroendocrine carcinoma in a child: morphologic, immunocytochemical, and molecular biologic studies. Cancer 71:2660–2665.

13- Warner TFCS, Insook S, Madura JA, Polak JM, Pearse AGE.(1980) Pancreatic

Polypeptide-producing apudoma of the liver. Cancer 46: 1146–1151.

14- Gary Schwartz, Agnes Colanta, Harold Gaetz, John Olichney, Fadi Attiyeh.

(2008) Case report: Primary carcinoid tumors of the liver. World Journal of

Surgical Oncology 6:91 doi: 10.1186/1477-7819-6-91.