

Primary Hepatic Carcinoid : A Case Report

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Abstract : Carcinoid Tumors are most common neuroendocrine tumors of gastrointestinal tracts. Carcinoid tumors of liver most commonly are metastatic. Primary hepatic carcinoid tumors are extremely rare neoplasms derived from hormone-producing neuroendocrine cells and first case was documented by EDMONSTON 1958. It is difficult to make their diagnosis before biopsy, surgical resection or necropsy. Only 60 cases were reported till 2008 and recent publication reported 94 cases. The most effective treatment is hepatectomy, and resection is determined by size and location of the lesion.

Introduction

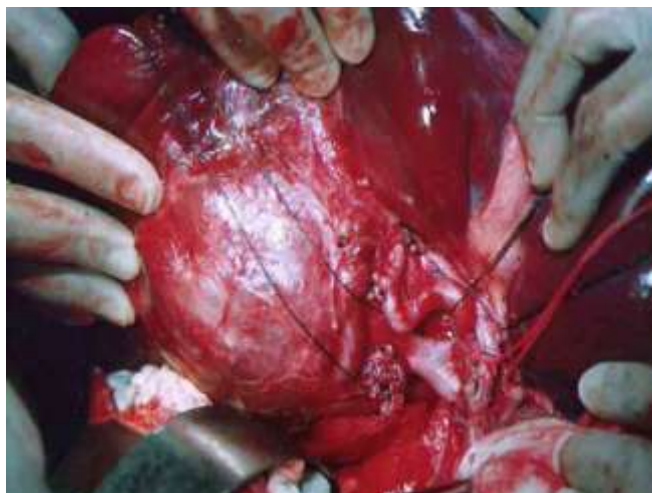
Primary Hepatic carcinoid are thought to arise from neuroendocrine cells (Kulshitzsky cells) located in the intrahepatic biliary epithelium. Chronic inflammation of the bile canaliculi can cause intestinal metaplasia, which predisposes the development of neuroendocrine tumors. The primary hepatic carcinoid tumor has no sex predilection and has no association with cirrhosis or preexisting liver disease. It accounts for only 0.3% of the cases of neuroendocrine tumors.¹ The diagnosis may be difficult preoperatively based on Imaging techniques. Some cases show association with HBV and HCV and on CT show findings of arterial hyperenhancement with venous washout suggestive of H.C.C, so without a biopsy or resection diagnosis is difficult and it may often be confused with Hepatocellular carcinoma particularly in areas with high HCC prevalence. CECT abdomen and MRI with gadolinium contrast will demonstrate tumor hypervascularity which can be further corroborated with Angiography. Primary hepatic carcinoids presents as a solitary tumor, with occasional extrahepatic metastasis; related to size >5cm with lung lymph nodes and bone being common sites. Octerotide scintigraphy can help delineate both primary as well as extrahepatic metastasis. Immunohistochemistry shows Chromogranin A positive in >80% cases and also 5HIAA positive in majority of cases.²⁻⁵

Case report

A 30 year-old female presented with gradually progressive abdominal lump in right hypochondriac region over one year. The laboratory test results showed normal complete blood count and liver function tests, negative serology for hepatitis, tumor markers (AFP & CEA) within normalcy. Clinical examination revealed massive hepatomegaly and ultrasound revealed SOL Right lobe of liver of approximately 8x6 cm²; CBD, CHD & IHBR were normal. CT scan confirmed the sonographic findings. The patient was explored via a Modified chevron incision and intraoperative findings revealed a large hypervascular solid mass lesion involving the right lobe of liver. An extended right hepatectomy was performed. The preoperative and postoperative course of patient was uneventful. Patient was discharged on 10th POD. The histopathological examination confirmed the diagnosis.



Right lobe liver mass with extensive hepatomegaly



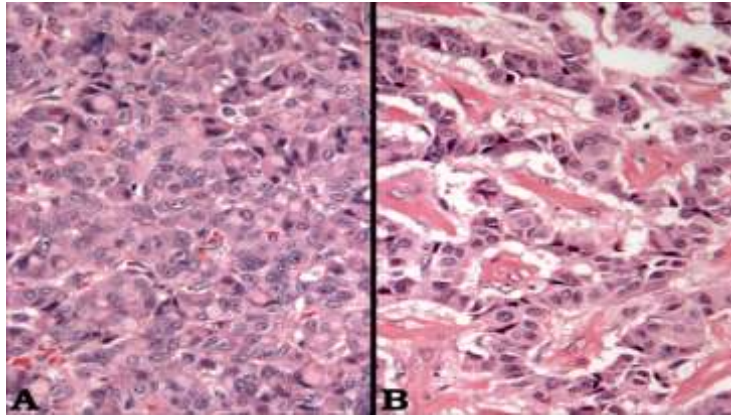
Surgery in progress- Right hepatic artery being ligated



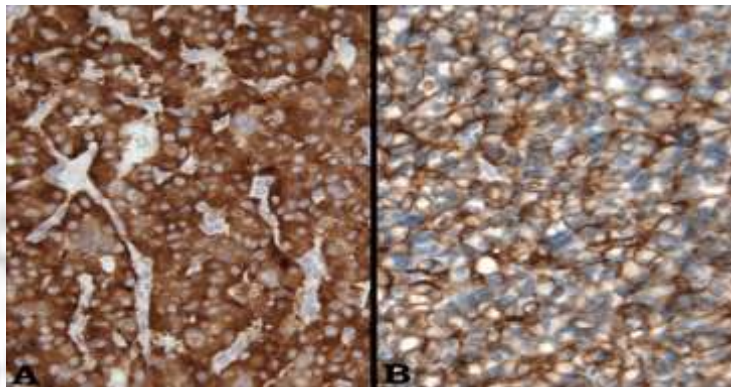
Resected Specimen: 7 x 5 cm² solid cystic mass lesion.



Post Operative CT-scan



Microscopic image of the specimen



Immunohistochemistry was positive for synaptophysin and CD56

Discussion

Primary hepatic carcinoid tumors may be an incidental finding via USG or CECT abdomen performed frequently for pain abdomen. Some patients present with severe symptoms including abdominal pain, jaundice, palpable right upper quadrant mass, carcinoid syndrome, carcinoid heart disease, and Cushing's Syndrome. Less than 10% of gastrointestinal carcinoids present with the carcinoid syndrome and when the syndrome is present it is almost always associated with hepatic metastasis. The Carcinoid Syndrome is rarely present in PHCT, with only two reported cases till date.⁴ The rarity of PHCT makes it difficult to diagnose accurately before biopsy or resection. The accurate diagnosis of carcinoid tumor before operation is very important because it suggests investigations to seek possible sites of metastasis; avoiding unnecessary operation or a second operation if a metastatic lesion is found. Imaging studies of any hepatic mass should begin with ultrasound and a triple-phase CT scan. MRI is increasingly being used, with improved visualization of carcinoid tumors on T2-weighted images. Additional information can be gained from nuclear medicine imaging scans, specifically utilizing Technetium-99m isotopes and octreotide scintigraphy. If carcinoid is diagnosed postoperatively on histopathology, upper and lower gastrointestinal endoscopy must be performed to look for primary GI carcinoid. After the appropriate workup of a hepatic mass, initial management is surgical resection when possible. Extent of resection is determined by location and size of the tumor(s), with multicentric bilobar disease often precluding resection, in which case alternative therapies include radiofrequency ablation (RFA), hepatectomy with OLT (transplantation), selective hepatic artery embolization (HAE), Portal vein chemoembolization (PACE) & intravenous octreotide infusion for symptomatic relief.¹⁻³

Conclusion

Metastatic liver involvement in gastrointestinal carcinoid is common more so if it is associated with carcinoid syndrome. However, the primary involvement of this organ is very rare. Better diagnostic methods are required to make a more precise preoperative differentiation between primary hepatic carcinoid tumor and hepatocellular carcinoma. Primary hepatic carcinoid tumor should be suspected in patients with no chronic liver disease, normal alpha-fetoprotein levels and solid-cystic lesion in imaging tests. The management is Hepatectomy determined by local extent of tumor whereas advanced cases need palliative treatment.

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