CASE REPORTS

PRIMARY NEUROENDOCRINE TUMOUR OF LIVER

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ABSTRACT

Primary neuroendocrine tumors of the liver are seldom seen. This is the case report of 45 years old woman who presented with right upper quadrant vague abdominal pain and low grade fever. On examination a firm mass about 6 × 4 cm was palpable in left hypochondrium and epigastrium. Ultrasound and CT-scan showed mass of left lobe (segment II and III) of liver. There was no evidence of cirrhosis and hepatitis B and C status was negative. The tumor markers were also within normal values. Core biopsy confirmed a tumor of neuroendocrine origin. Octreotide scan was done which confirmed that liver was the primary site of neuroendocrine tumor. Left lobectomy (Segment II & III) was done. The diagnosis was confirmed on histopathology and immune histo-chemistry of specimen.

Keywords: Immunohistochemistry, Neuroendocrine carcinomas, Octreotide.

INTRODUCTION

Liver is the most common site of metastasis for neuroendocrine tumors of gastrointestinal tract[1]. Primary hepatic neuroendocrine carcinomas (PHNECs) are extremely rare, with only about 150 cases having been reported in the literature[2]. The diagnosis of PHNEC requires strict exclusion of possible extra-hepatic primary sites for its diagnosis[3,4]. Neuroendocrine tumors typically produce hormones and or amines. About one-half of all neuroendocrine tumors are described as non-functioning[5]. Neuroendocrine tumors are diagnosed by pathologic confirmation and may demonstrate insular, trabecular or glandular cell arrangement[2,6]. Immuno-histo-chemical staining shows positive immunore-activity to chromogranin A, neuron specific enolase, synaptophysin, and CEA[2,6].

CASE REPORT

A female aged 45 years resident of Khyber agency presented to Military Hospital Rawalpindi surgical OPD with complaints of pain right upper quadrant of abdomen, nausea and fever for three months. The pain was dull, aching in nature, gradual in onset, mild in intensity with no radiation. There was associated nausea and low grade fever along with body aches, weakness and no weight loss. There was no history of jaundice or previous abdominal surgery. Rest of the past, personal, family, drug and socioeconomic history were not contributory. On abdominal examination there was mild tenderness in the right hypochondrium with a

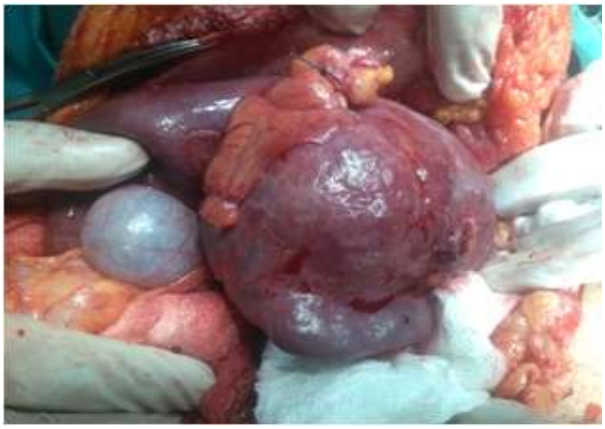

Figure: Per-operative view of tumor in left lobe of liver.

mass about 5 × 6 cm in size, palpable indistinctly and the liver edge palpable about 2 cm below costal margin. There was no jaundice, ascites and anemia. Investigations including blood complete picture, urea, creatinine, liver function tests, PT/APTT, viral markers and chest x-ray were done which were within normal limits.

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Tumor markers including Alpha Fetoprotein, CEA and CA19-9 were also normal. CT scan showed large cystic/solid mass approximately 6x8x7 cm in segment III of liver with hepatosplenomegaly. Percutaneous Core-Cut biopsy was done which revealed neuroendocrine tumor. Based on biopsy report, octreotide scan was done to locate the primary lesion and it showed that the tumor was only present in left lobe of liver (segment II and III). Immuno-histochemistry was done on the biopsy sample subsequently and positive reactivity to chromogranin and synaptophysin confirmed neuroendocrine origin, and negative reactivity to CDX 2, PAX 8 and TTF 1 favored primary hepatic origin. So, a final diagnosis of Primary neuroendocrine tumor of liver was made.

The patient was prepared for surgery and laparotomy was performed through the J-shaped incision and per operatively tumor was about 7×7×8 cm solid/cystic in consistency and mainly involving liver segment III and part of segment II, adherent to lesser curvature of stomach (figure). Left lobectomy was done by ligating inflow and outflow of both segments. Abdomen was closed with drain in place. Post operatively the patient was nursed in HDU and the recovery was uneventful. The patient was discharged after 01 week and histopathology specimen showed moderately differentiated Primary Hepatic neuroendocrine Carcinoma PHNEC with clear margins. The patient is on regular follow up on six monthly basis and has not shown any signs of recurrence.

DISCUSSION

Although there is a steady increase in the number of cases of PHNEC but this disease is still a rarity[7]. The meta analysis of reported cases so far shows that the disease is common between age of 37 to 80 years with median age of 66.5 with equal gender disposition[2]. Our case was a 45 years old female. Most of the patients presented with upper abdominal pain, weight loss, fatigue, fever and hepatomegaly with clinical manifestations of hormone secretion in less than 20%[7]. In our case the presentation was typical except there was no hormonal manifestation. Diagnosis of this problem is still a dilemma and utmost endeavors are made from pre-operative work up to post-operative histology to find the extra-hepatic primary. Pre-operative work up is aimed to locate the primary tumor and for this purpose CT-Scan, MRI, Octreotide scan and PET scan are implicated. Octreotide Scan is most efficient in detecting PHNEC and Donadon et al reported 88% specificity and 100% positive predictive value of octreotide scan in detecting PHNECs[7]. Other markers as AFP, CEA etc are nonspecific and are used to rule out alternative diagnosis. In our case octreotide scan confirmed the primary lesion in liver. The confirmation of PHNECs is based on histopathology and positive immuno-histochemistry for chromogranin and synapto-physin[2]; both found positive in our patient. The mainstay of treatment is surgical resection of tumor, Park et al reported 78% 5-year survival in his series of 48 patients with recurrence of 19%[2]. Aggressive hepatic resection is safe and in our case left lobectomy was performed. The chemo-therapy in PHNECs is still controversial because of insufficient data but there are cases where control and down staging is reported with octreotide analogs[2,7].

PHNEC is a very rare tumor which is often regarded as a metastatic lesion with primary elsewhere and diagnosed on histopathology and immunohistochemistry. Latest nuclear/ radiological scans have a role in detecting the primary neuroendocrine tumor of liver after exclusion.

CONFLICT OF INTEREST

This study has no conflict of interest to declare by any author.

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