CASE REPORT

Fibrolamellar Hepatocellular Carcinoma – A Rare Clinical Variant

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ABSTRACT

Fibrolamellar type is a rare variant of hepatocellular carcinoma (FLHCC). A 20 year old boy presented with pain and mass in the epigastric region for the last six months. Early diagnosis and confinement of the lesion to a lobe made it possible to perform complete resection of tumour along with subtotal gastrectomy. Patient recovered well and was sent to oncologist for further treatment.

Key words *Fibrolamellar hepatocellular carcinoma, Resection, Variant.*

INTRODUCTION:

Hepatocellular carcinoma (HCC) constitutes approximately 90% of the primary liver cancers in the United States. Microscopically there are four variants of HCC, which are fibrolamellar, pseudoglandular (adenoid), pleomorpic (giant cells) and clear cell type.[1] Fibrolamellar type is one of the rare clinical variants. NF-KB activation is involved in the genesis of fibrolamellar hepatocellular carcinoma (FLHCC). It represents a novel target for therapeutic intervention.[2]

The tumour does not produce alpha feto protein (AFP) and is associated with high level of neurotensin and vitamin B12 binding globulin.[3] The purpose of this case report is to highlight the clinical presentation, diagnosis and management of fibrolamellar HCC.

CASE REPORT:

A 20 year old boy resident of the neighbouring country presented with the complaint of mass and pain in the epigastric region for the last six months. Swelling gradually increased in size. Patient developed mild pain which was dull in nature. Patient also had dyspepsia. There was no history of weight loss, alcoholism and hepatitis. General physical examination revealed a medium built boy weighing 55Kg. There was no pallor, jaundice, cyanosis and lymphadenopathy. Systemic abdominal examination revealed mild tenderness in the epigastric region. A mass was found in the epigastric region measuring 9cm × 8cm. It was firm in consistency with non fixation to surrounding structures. No other abnormality was detected.

Biochemistry showed normal CBC. Total bilirubin was 0.9mg/dl, ALT 24 u/l, alkaline phosphatase 194 u/l. HBsAg and anti- HCV were non reactive. Serum alpha feto protein was 1.54 IU/l (normal range for adult is < 5.8 IU/l. Urinalysis was normal. Sonographic study revealed a 10.3cm×9.7cm, solid heterogeneous mass in the epigastric region arising from the left lobe of the liver, however liver span was 13.8 cm (normal size). No dilatation of biliary duct and portal vein was seen. Rest of the sonography of abdomen was normal. His contrast CT scan revealed a large heterogeneously enhancing solid mass (measuring 10cm×10cm) arising from left lobe of the liver reaching up to the lesser curvature of the stomach. Rest of the liver was without any findings (Fig I). Scan of the remaining abdomen was normal. His X-ray chest was reported as normal.

Fine needle aspiration cytology report showed scattered atypical polygonal cells with high N/C ratio, nucleoli and moderate eosinophilic neoplasm. Few oval to spindle shaped cells also seen. The diagnosis of fibrolamellar hepatocellular carcinoma was made.

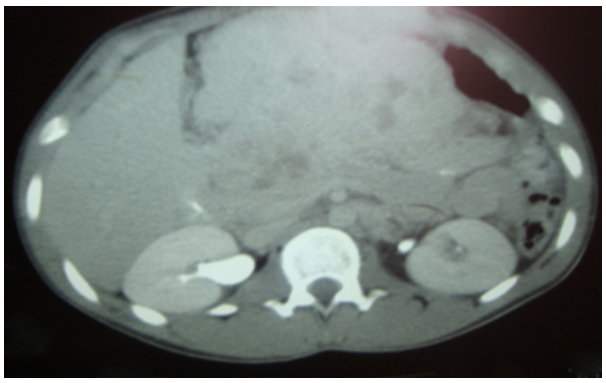

Fig I: CT Scan showing tumour in left lobe of liver

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Patient was informed about the diagnosis. A plan of surgery was made. Abdomen was opened via upper mid line incision. There was a mass attached on the under surface of left lobe of liver, involving the lesser omentum and lesser curvature of the stomach. Right lobe of the liver was not involved. There was no involvement of the caeliac lymph nodes. The mass was not fixed with underlying structures. Mass was resected from the liver by excising the under surface of liver. Subtotal gastrectomy was also performed. Gastrojejunostomy along with enteroentrostomy were also done. Histopathological examination showed fibrolamellar hepatocellular carcinoma. Resected specimen of stomach revealed involvement of the serosa at lesser curvature only, while rest of the stomach was normal. Patient was sent to oncologist for further management.

DISCUSSION:

Fibrolamellar hepatocellular carcinoma (FLHCC) is listed as a rare disease. In a study conducted by El- Serag HB, 71 patients diagnosed with fibrolamellar hepatocellular carcinoma out of 9870 patients with hepatocellular carcinoma.[4] Yoshimi F et al in a study conducted in Japan reported 18 cases of FLHCC up until 1999.[5] In another study conducted in Sweden University three patients were diagnosed as a case of FLHCC over a span of 22 years.[6]

Yen JB, reported a case of fibrolamellar hepatocellular carcinoma in a 14 years old girl. The mass was about 10 cm in size in the left lobe of liver.[7] Our patient was a 20 year old boy. In a study conducted by Almani SA, et al sensitivity of alpha-fetoprotein in hepatocellular carcinoma was 72%, specificity 89%, positive predictive value 86.7% and negative predictive value of 76.1%.[8] In our patient alpha feto protein was within normal limit. Biphasic contrast enhanced helical CT is a useful method in detection and characterization of HCC. CT scan of fibrolamellar hepatocellular carcinoma demonstrates a large heterogeneously enhancing mass with a hypo attenuating central scar and punctuate central calcification.[9] Same were the findings in reported patient. Imaging studies have a major role in clinical diagnosis but pathology is the gold standard in confirming diagnosis. Characteristic features were noted in our patient based upon which final diagnosis was made.[10] The overall 1- year relative survival rates were significantly longer in patients with FLHCC than HCC (73.3% in patients with FLHCC and 26.0% in HCC.[11] At three months follow up our patient is doing well.

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