

## Case Report

### A Case Report of Fibrolamellar Hepatocellular Carcinoma in a Young Adult

Muhammad Atiq<sup>1</sup>, Muhammad Haroon<sup>2</sup>, Muhammad Imran Khan<sup>3</sup>, Ahmed Siddique Ammar<sup>4</sup>, Warda Khalid<sup>5</sup>, Faisal Hanif<sup>6</sup>

<sup>1-6</sup>HPB & Liver Transplant Unit, Bahria International Hospital Orchard Lahore Pakistan

#### Abstract

Among the four histopathological variants of hepatocellular carcinoma, Fibrolamellar hepatocellular carcinoma (FLHCC) is a rare variant of hepatocellular carcinoma (HCC). It occurs in less than 1 % cases of hepatocellular carcinoma with distinct features as compared to classical HCC. It differs from typical HCC in terms of epidemiology, etiology, clinical presentation and prognosis. In this case report we are going to present a case of fibrolamellar variety of HCC in a 20-year young male with no history of previous liver disease and no characteristics features on Computed Tomography (CT) scan when compared with typical HCC. After discussing in multidisciplinary meeting, he underwent non anatomical liver resection which remained uneventful. Histopathology of the biopsy sample revealed FLHCC. No neoadjuvant therapy was given. Patient was kept on 1 year follow-up and no recurrence or metastasis occurred.

**Corresponding Author** | Dr. Ahmed Siddique Ammar, Senior Registrar, HPB & Liver Transplant Unit, Bahria International Hospital Orchard Lahore Pakistan **Email:** Asammar1912@gmail.com

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#### Introduction:

Hepatocellular carcinoma (HCC) is the sixth most common cancer worldwide and constitutes about 90% of all primary liver tumors. There are four histopathological variants of HCC which include adenoid, giant cells, fibrolamellar, and clear cell type<sup>1</sup>.

Fibrolamellar hepatocellular carcinoma (FLHCC) is a rare variant of HCC which represents only 1% of all primary hepatic tumors. It is different from classical HCC in etiology, clinical and radiological presentation<sup>2</sup>. As there are no usual risk factors such as Hepatitis B, C or cirrhosis and radiological findings can be atypical so it could be difficult to diagnose and cause delay in treatment<sup>3</sup>.

In this case report we going to present a case of rare variety of HCC known as Fibrolamellar hepatocellular carcinoma (FLHCC) in a 20-year-old male. The purpose of this case report is to spread awareness to the readers about presentation, management plan and prognosis of FLHCC.

#### Case Report:

A 20 years old male with no comorbidities, presented to outpatient department of Bahria International Hospital Orchard, Lahore Pakistan in June 2020 with complaint of right hypochondrium pain for two months. The pain was mild in intensity, dull, non-radiating. He had no history of jaundice, hematemesis, fever, and anorexia or weight loss. Viral markers were negative. No abnormal findings were appreciated on abdominal examination. All baseline blood reports were within normal range, and the level of alpha fetoprotein (AFP) was 2.1 IU.

An ultrasound (USG) abdomen was performed which showed 57 x 38 x 31 mm hypoechoic mass in segment 5 of liver. There were no liver parenchymal changes, splenomegaly or ascites. Computed tomography (CT) Triphasic abdomen scan showed a heterogeneous lesion in segment 5 and 4b of liver (Figure 1). This lesion showed patchy arterial enhancement with the impression of washout on portal venous and delayed phase suggestive of liver adenoma. There were no

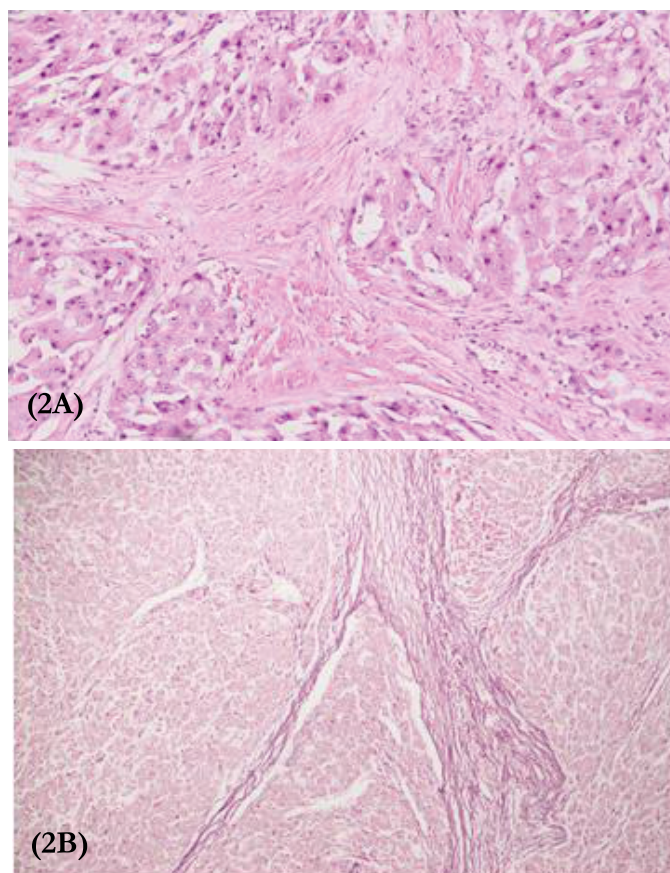


**Figure 1:** Contrast enhanced computed tomography (CECT). The lesion in the segment IV b and V. 1A: White circle showing arterial phase showing Patchy arterial enhancement. 1B: Red Circle Showing venous phase with gradual washout of contrast medium.

enlarged or suspicious lymph nodes and no visceral and peritoneal abnormality.

The case was discussed in multidisciplinary meeting (MDM) and the recommendation was to proceed with liver resection. A written informed consent was taken from the patient for operation under general anesthesia. Non anatomical liver resection was performed and the surgery remained uneventful. Post operative period was symptom free and he was discharged on seventh post operative day. Later histopathology showed FLHCC (Figure 2). Tumor size was 2.1 cm with well-preserved margins. All 5/5 lymph nodes were negative for HCC. The pathological stage was T1bN0.

Staging CT scan of chest was done in the follow up period which showed no metastasis. Final stage of the



**Figure 2:** Histopathological findings for FLHCC. 2A: Nests of neoplastic hepatocytes separated by fibrous bands of connective tissue. Large, polygonal hepatocytes with ample eosinophilic cytoplasm, slight nuclear pleomorphism and evident nucleoli can be seen. 2B: Stain highlights the bands of fibrous connective tissue that separate the nests of neoplastic hepatocytes.

tumor was pT1bN0M0. No adjuvant treatment was offered. Neither recurrence nor any metastases was observed on follow-up CT scan at 1 year. Patient is kept on surveillance with liver dynamic CT scan.

#### Discussion:

FLHCC was first described in 1956 by Admondson and initially it was considered as a different identity. It represents up to 1% of all the primary tumors of the liver. FLHCC typically affects the younger adults with median age of 25 years with equal gender predominance<sup>4</sup>. As compared to typical HCC, causative factors for FLHCC are unclear. It commonly develops on the background of normal hepatic parenchyma with no history of viral hepatitis or metabolic abnormalities<sup>5</sup>.

On gross anatomy, FLHCC presents with encapsulated heterogeneous mass with areas of necrosis and increased vascularity. It has many fibrous septa which connect

to central lesion similar to Focal Nodular Hyperplasia (FNH) causing confusion in diagnosis. Microscopically, these cancers have large polygonal cells with macro nucleoli and abundant fibrous stroma which is responsible for its name fibro lamellar<sup>6</sup>.

FLHCC normally presents with non-specific symptoms and signs like abdominal pain, malaise and anorexia or weight loss. Laboratory findings are usually normal in FLHCC including AFP levels. Classic CT findings of HCC include a hyper vascular pattern with arterial enhancement and rapid washout during the portal venous phase<sup>7</sup>. CT scan without Contrast FLHCC usually present as big, isolated hypoattenuating mass which is surrounded by well-defined wall and its surface is lobulated. However, in triphasic CT scan, during the arterial phase this variety of HCC become more enhanced and hyperattenuating. Similarly, during portal and delayed phases of triphasic CT the density of this particular lesion becomes more similar to the surrounding normal liver parenchyma<sup>8</sup>. During the portal and delayed phases, the tumor remains enhancing and becomes more homogeneous in appearance, with its density more closely matching that of the liver as equilibrium is achieved. These findings are similar to CT Triphasic abdomen done in our case which also showed patchy arterial enhancement with the impression of washout on portal venous and delayed phase. The ideal treatment for these patients is surgical resection. The role of chemotherapy and sorafenib is not clear as there are no specific clinical trials for this variant<sup>9</sup>. Prognosis of FHCC is also uncertain. Initially FHCC was presumed to have better survival rate due to absence of cirrhosis but some recent studies had showed that its prognosis is not as favorable as it was considered. A study by Walsh et al showed that after liver resection 1-, 3- and 5-years survival rates were 93%, 80% and 70% respectively<sup>10</sup>.

### Conclusion:

FLHCC is rare variant of typical HCC which should be suspected in young patients with no underlying liver disease or cirrhosis. In cases where patient is young with no co morbidities and liver is otherwise normal the biopsy of localized lesion must be done. Surgery is the treatment of choice for FHCC where possible.

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