CASE REPORT

doi: 10.5455/medarh.2022.76.387-390
MED ARCH. 2022 OCT; 76(5): 387-390
RECEIVED: SEP 14, 2022
ACCEPTED: OCT 22, 2022

Case Report of Fibrolamellar Hepatocellular Carcinoma in A 15-Year-old Male

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ABSTRACT

Background: A rare form of hepatocellular cancer is called fibrolamellar hepatocellular carcinoma (FL-HCC) which occurs mostly in young adults who are medically free, regardless of their gender. It usually presents with abdominal pain with right upper quadrant palpable mass, nausea, and weight loss associated with higher Alpha-Fetoprotein (AFP) in some cases. **Objective:** We report a case of a 15-year-old male patient who was diagnosed with (FL-HCC), successfully treated with surgical resection and is currently free of relapses. **Case presentation:** A 15-year-old male patient with no previous medical or surgical history, presented with recurrent vomiting for two months, weight loss, and loss of appetite. Patient presented with normal systemic examination except for abdominal examination which revealed a generalized distended abdomen with mild tenderness in the right upper quadrant with the presence of hepatomegaly. Laboratory and radiological investigation showed high level of (AFP). CT and liver MRI showed large right hepatic lobe lesion then TRU-CUT needle biopsy was performed which showed Fibrolamellar hepatocellular carcinoma and patient underwent surgical resection with no postoperative complication followed by multiple cycle of chemotherapy and no signs of relapse with 3 year follow up. **Conclusion:** Fibrolamellar hepatocellular carcinoma is rear type hepatocellular carcinoma which occurs mostly in young adults who are medically free with vague symptom and to diagnose it need high index of suspicion and variers Laboratory and radiological investigation including biopsy. However, it can be treated successfully by surgical resection followed by chemotherapy in selected cases if diagnosis in timely manner.

Keywords: Hepatocellular cancer, Fibrolamellar Hepatocellular Carcinoma (FL-HCC).

1. BACKGROUND

An uncommon primary hepatic malignancy is fibrolamellar hepatocellular carcinoma (FL-HCC). Concerning its behaviour and clinical signs, it differs from conventional hepatocellular carcinoma (HCC) and distinctively, it occurs in young adults and teens without underlying liver pathology. Fl-HCC is responsible for one percent of primary liver cancer cases (1). We present a case of a 15-year-old male patient who was diagnosed with (FL-HCC), successfully treated with surgical resection and is currently free of relapses.

2. OBJECTIVE

We report a case of a 15-year-old male patient who was diagnosed with (FL-HCC), successfully treated with surgical resection and is currently free of relapses.

3. CASE PRESENTATION

A 15-year-old male patient with no previous medical or surgical history , came to the clinic complaining of recurrent vomiting for two months, weight loss, and loss of appetite. The patient has no history of jaundice, changes in the bowel habits, or urinary symptoms. Also, the patient has no family history of any type of malignancy.

Vital signs were within the normal range. Patient presented with normal systemic examination except for abdominal examination which revealed a generalized distended abdomen with mild tenderness in the right upper quadrant with the presence of hepatomegaly.

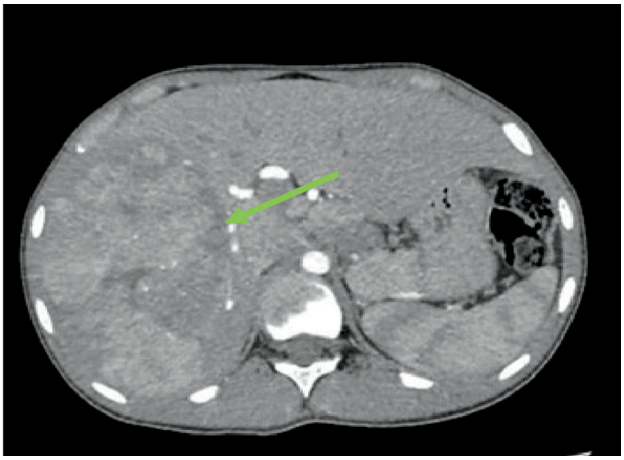

Figure 1. Axial view of a CT liver showing the green arow indicating large solitary right hepatic lesion in the liver with central scar and calcification.

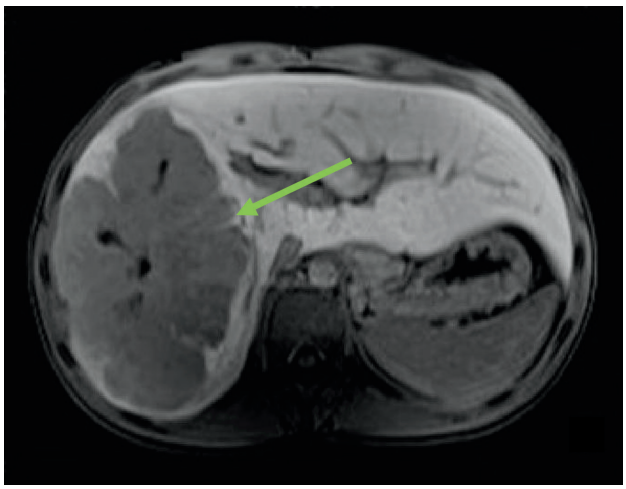

Figure 2. Axial Axial view of liver MRI show the green arow indicating large mass in right lobe with central scar.

Our workup included complete blood count (CBC) and liver function tests (LFT) which were within normal ranges. Tumour markers were ordered, Alpha Fetoprotein was 50.8 ng/ml (1.09-8.04), which is markedly elevated. Meanwhile, Cancer Antigen 19-9 (CA 19.9) and carcinoembryonic antigen (CEA) were within normal ranges ((CA 19.9 10.76 (0-37)) and (CEA > 0.73 (0-5)).

With intravenous (IV) contrast, we performed a computed tomography (CT) scan of the chest, abdomen and pelvis which showed smooth liver contour without signs of cirrhosis with large right hepatic lobe lesion occupying almost the whole segment with heterogenous arterial enhancement and iso-enhancement in the port venous phase without significant washout measuring 13.1 x 9.2 x 13.3 cm in AP x t x CC dimensions as shown by (Figure 1).

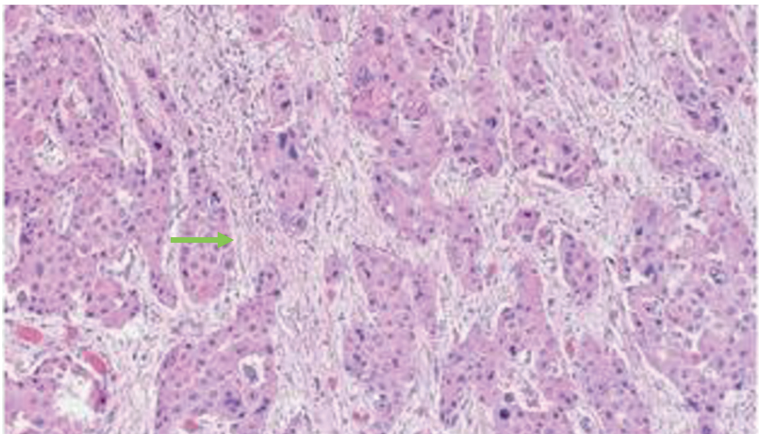

Figure 3. Histopathological view of the tumour show the green arow indicating neoplastic hepatocytes separated by fibrous bands of connective tissue.

There was also a hypodense centre with multiple foci of calcifications that displaces the right hepatic artery and vein without thrombus. There were lymph nodes involvement in porta hepatis and gastro-hepatic groups, the largest measurement was 2.9 x 1.9 cm. No significant retroperitoneal lymph nodes. There are no metastases were detected in the exam.

Liver MRI was done and showed Large right hepatic lesion show isointense T2 and low signal intensity on T1 with central hypointense T1/T2 with multiple foci of calcification and no diffusion restriction or fat component. measuring 13.1 x 9.2 x 13.3 cm in AP x t x CC dimensions. There were lymph nodes involvement in porta hepatis and gastro-hepatic groups, the largest measurement was 2.9 x 1.9 cm. No significant retroperitoneal lymph nodes (Figure 2).

After 5 days, the patient underwent an ultrasound guided TRU-CUT needle biopsy which showed Fibrolamellar hepatocellular carcinoma as shown by (Figure 3). Then, the case was discussed in the tumour board and the decision for a surgical resection was made.

The patient was admitted after 10 days from the tumour board meeting for resection. He underwent formal right hepatectomy with portal lymph node excision and open cholecystectomy. The postoperative hospital stay was uneventful, and the patient was discharged on postoperative day 8 in good condition. A histopathological review of the specimen showed well differentiated multifocal fibrolamellar hepatocellular carcinoma with 1.4cm free margin . there is evidence of lymphovascular invasion in small vessel - the pathological stage is pT3,pN1.

The patient was followed in the outpatient clinic in good condition.. Then, our patient was referred to Medical oncology and chemotherapy was offered for the patient but patient refused. After 1 year patient return to our outpatient clinic and patient was revaluated and a positron emission tomography (PET) scan was done showed no singe of metastasis and offer chemotherapy aging and the patient accept and receive 2 cycles of cisplatin and Taxotere then 3 cycle of cisplatin and tacol then due to national shortage of this type of chemotherapy change to docetaxel and carboplatin for 3 cycle. and the patient is still following up with oncology team in good condition for 3 year.

4. DISCUSSION

With an incidence rate of 0.02 per 100,000, fibrolamellar hepatocellular carcinoma (FL-HCC) is an uncommon form of hepatocellular carcinoma that accounts for 0.85% of all primary liver cancer cases (2). According to the Surveillance, Epidemiology, and End Results (SEER) database, the average age at presentation is 25.6 12.6 years, with a significant increase in late adolescence and a peak in the early twenties (3).

It typically does not occur in conjunction with any known chronic liver diseases (4). Weight loss (60%), abdominal mass (50%), and abdominal discomfort (65%) are the most common symptoms in patients with FL-HCC (5). The majority of patients in radiological investigations (80%) had a single nodule, and the median tumor size is 120 mm (6.6-19 cm) (5).

For our case, the patient presented with recurrent vomiting for two months, weight loss, and anorexia and he had an abdominal mass measuring 13.1 x 9.2 x 13.3 cm in Anterior-posterior (AP) x transverse (t) x Craniocaudal (CC) dimensions, respectively, with a large multiple porta hepatis and gastro-hepatic lymph nodes, the largest measurement was 2.9 x 1.9 cm. Aspartate aminotransferase (AST) and alanine aminotransferase (ALT) serum levels can be normal or modestly increased in FLHCC (66). Alkaline phosphatase levels could also be increased (7). Other elevated serumproteins include fibrinogen (8). Some immunohistochemical abnormalities help distinguishing (FL-HCC) from conventional hepatocellular carcinoma (HCC), such as low AFP, high serum unsaturated vitamin B12 binding capacity, and elevated serum neurotensin (7).

Our patient had Elevated Alpha Fetoprotein > 50.8 ng/ml (1.09-8.04), other labs were within normal ranges. A definitive diagnosis can be made by biopsy whether Fine needle aspiration (FNA) or trucut (TRU-CUT) biopsy (9). A TRU-CUT biopsy was taken in our case. Diagnostic images are helpful in the diagnosis of (FL-HCC). The most frequent finding is hepatomegaly, and tumor calcification is frequently seen. (FL-HCC) presents on Ultrasound (US) as a single, well-defined lobulated mass with varied echotexture, composed of hyperechoic or isoechoic components (10). The central scar, which presents in 33%- 60% of patients, may be visualized as a central area of hyper echogenicity (11). Computed Tomography (CT) presentation is usually a solitary, hypo-attenuating mass with a well-defined lobulated surface (4). The central scar is not increased, making it possible to tell the fibrolamellar form of hepatocellular carcinoma apart from lesions that develop later, including focal nodular hyperplasia (FNH) (13). In 33%-58% of (FL-HCC) patients, there is calcification inside the core scars which is consistent with our case (7). (FL- HCC) presents as a large, firm, grey, solitary mass, as opposed to HCC, which is frequently numerous, soft, and hemorrhagic (8). (FL-HCC) frequently manifests in a noncirrhotic liver as a well-circumscribed tumor with a conspicuous fibrous tissue and core stellate scar, resembling focal nodular hyperplasia (FNH) (12). Histologically, the tumor is composed of massive polygonal cells with lamellar fibrosis, huge vesiculated nuclei, and big nucleoli (6). Patients with HCC have a resectability rate of 10 to 23%, compared to 48 to 100% for (FL-HCC) described in the literature (8). Which was the chosen treatment for this case with Portal lymph node excision and open cholecystectomy. Patients with (FL- HCC) had a relative one-year survival rate of 73.3%.

5. CONCLUSION

(FL-HCC) is a unique liver malignancy at clinical, histological, and molecular levels. It tends to occur in young individuals. Its etiology is unknown and its molecular aspect is poorly understood. However, (FL-HCC) can be treated successfully by surgical resection followed by chemotherapy in selected cases. Although (FL-HCC) has been reported from a many countries besides Saudi Arabia, recent data and studies are still required to understand the nature of this tumor and to improve the overall survival rates.

- **Ethics approval and consent to participate**: Ethical approval is not required at our institution to publish an anonymous case report and written inform consent was obtain from the patient parent.
- **Consent for publication**: Informed consent was taken for publication of this paper.
- **Availability of data and materials**: Not applicable.
- **Authors' contributions**: HA, RA, MM, AA, RZ were all major contributors in writing the manuscript. HA, HA, AA performed the surgery and contributed by writing their intraoperative findings in the manuscript along with the intraoperative images. All authors read and approved the final manuscript.
- **Conflict of interest**: The authors declare that they have no conflict of interest.
- **Financial support and sponsorship**: None.

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