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Case Report
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De novo Juvenile Hepatocellular Adenocarcinoma

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Abstract

Hepatocellular adenocarcinoma is primary liver cancer which commonly arises in cirrhotic liver but it can arise *de novo*. This is a case of 14 years old boy who had no underlying liver disease, presented first time with right hypochondriac pain and documented fever for 1 month. On examination he had tender hepatomegaly with liver span of 16 cm. His initial ultrasound abdomen given impression of large cavernous hemangioma so triphasic CT scan abdomen was performed which revealed 10 cm hepatoma in right lobe of liver with few other small lesions. Serum alpha fetoprotein was 313.46 ng/ml. He was then referred to surgical team and his tumor was completely resected and excisional biopsy confirmed the diagnosis of HCC.

Keywords: Hepatocellular adenocarcinoma; Juvenile; *De novo*

Introduction

Hepatocellular adenocarcinoma is a malignant primary tumor of liver [1]. Worldwide, in adult population it is reported to be 5th & 7th commonest cancer in males and females respectively [2]. It commonly arises on background of cirrhosis irrespective of the cause of cirrhosis and chronic viral hepatitis B and C [3].

In children, hepatocellular adenocarcinoma is a rare malignancy [4]. *De novo* multicentric hepatocellular adenocarcinoma is even more rare in young population. It occurs in teenagers usually with underlying chronic liver disease and cirrhosis [5]. There are studies which are showing occurrence of *de novo* hepatocellular carcinoma in adult patients with chronic viral hepatitis who do not have developed cirrhosis and also among post liver transplant patients [6].

Fibrolamellar carcinoma is a subtype of hepatocellular carcinoma occurs in childhood [7], like Hepatocellular adenocarcinoma it arises in the background of chronic liver disease due to any cause like glycogen storage disease, chronic viral hepatitis and cirrhosis as well as some times it arises *de novo* [8].

Case Presentation

A 14 years old male with no known previous co-morbidities presented to our department with complaint of right upper quadrant pain and fever documented up to 101°F for 30 days. He didn't have significant past or family history of liver diseases. On physical examination patient was of average height and thin built who was vitally stable. Physical examination showed no stigmata of chronic liver disease. Abdominal examination showed mild tenderness in right hypochondrium, with palpable liver of 3 cm below right costal margin and liver span of 16 cm. There was no splenomegaly or signs of free fluid seen. CBC showed hemoglobin 12.9 g/dl, white cell count 14.6 × 10^9/L with 86% neutrophil count and platelets 258 × 10^9/L. LFTs showed total bilirubin of 1.5 mg/dl, alanine transferase 68 U/L, alkaline phosphatase 632 U/l, gamma glutamyltransferase 54 U/L, serum creatinine 0.59 mg/dl, serum albumin 3.7 g/dl, INR 1.22. HBsAg and anti-HCV were nonreactive. Serum alpha fetoprotein was 313.46 ng/ml.

His initial ultrasound abdomen gave impression of large cavernous hemangioma and repeat ultrasound abdomen revealed enlarged liver normal texture and homogeneous echogenicity. Few mass lesions were seen in right lobe of liver, the largest one is 10.6 cm × 9.8 cm in size and heterogeneous in echotexture. His triphasic CT scan abdomen revealed enlarged liver with regular margins. Few hypervascular lesions were seen in both lobes of liver predominantly involving segment IV, part of segment VIII and left lobe showing arterial enhancement (Figure 1a) and washout on delayed images (Figure 1b), suggestive of hepatoma. Largest lesion measured approximately 8.6 cm × 9.0 cm × 9.7 cm in AP × TS × CC dimensions. Other lesions seen in segment V measured approximately 4 cm × 4 cm and in segment II and III approximately 1.5 cm × 1.3 cm and 2.5 cm ×

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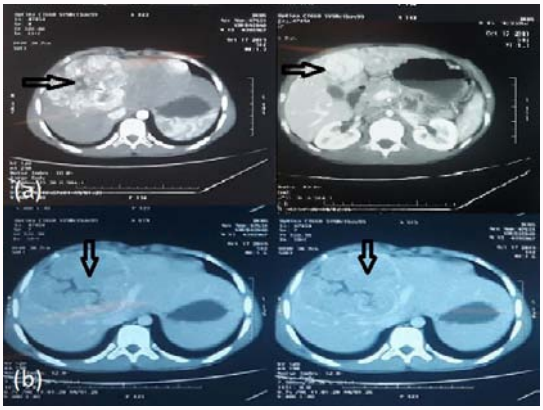

Figure 1a: CT scan abdomen triphasic axial sections showing an irregular mass on right lobe of liver also involving left lobe with diffuse enhancement on arterial phase (Right side) another small lesion is seen in segment V with arterial enhancement (left side) **(b)** delayed washout on portovenous phase(orange arrows).

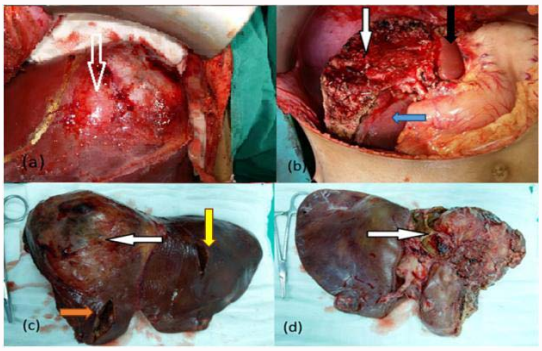

Figure 2: Operative Findings: (a) showing tumor on anterior surface predominantly segment IV and VIII, (b) liver bed (white arrow) after tumor resection showing preserved caudate lobe (black arrow) segment VI and VII (blue arrow). (c) Resected liver segments showing tumor anteriorly in segment IV and VIII (white arrow), another lesion on segment V (orange arrow)and segment II & III (yellow arrow)and posteriorly (d).

3 cm respectively. Thrombus was also seen in the lateral division of left branch of portal vein showing arterial enhancement and washout on delayed images, suggesting tumor thrombosis. Portal vein was normal in Calibre.

Treatment & Follow-up

The case was discussed in multi-disciplinary team meeting and decision was taken for tumor resection. Despite the large size and widespread lesion but since underlying liver was normal and not cirrhotic, resection was possible. Later his tumor was resected through extended left hepatectomy in which segment 2, 3, 4, 5 and 8 were resected and sent for histopathology.

Post-surgery patient's Serum alpha fetoprotein has declined to 8.04 ng/ml after two weeks which was favourable indicating complete tumor resection.

Histopathology report revealed malignant infiltrative neoplasm in sinusoidal and trabecular pattern with scattered chronic inflammatory cell infiltrate. Hilar vessels were tumor free. The tumor cells showed positive expression of Immunohistochemical stains Cytokeratin 8/18 and Hep Par 1 (Figure 3).

This was suggestive of moderately differentiated Hepatocellular

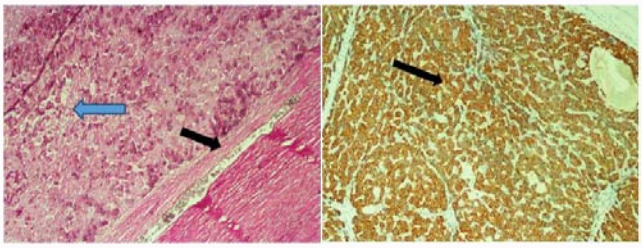

Figure 3(a): Excisional biopsy specimen of liver lesion showing encapsulated (black arrow) abnormal neoplastic cells (blue arrow). 2(b): Tumor cells are positive for Hep Par 1 Immunohistochemical stains (black arrow).

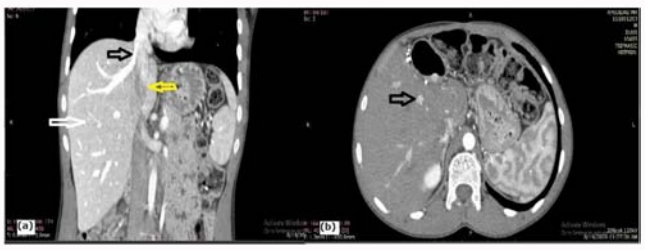

Figure 4: CT Triphasic (a) coronal section showing segment IV & VII (white arrow) and caudate lobe (yellow arrow) with right hepatic vein draining into inferior vena cava (black arrow), (b) axial section showing small arterially enhanced lesion in segment V (black arrow).

Adenocarcinoma with no lymphovascular invasion and there was no evidence of cirrhosis present in adjacent liver however moderate chronic inflammation noted.

This patient is in close follow-up and is doing well post-surgery. Patient has gained weight from 29 kg to 38 kg. After ten months repeat triphasic CT scan showed small lesion 0.7 cm × 0.4 cm in segment V with arterial enhancement but no definite washout on delayed phase. Serum Alpha fetoprotein declined to 3.1 ng/ml and LFTs become normal. This case again discussed in multidisciplinary team meeting and decided to keep on follow up with triphasic CT scan after 3 months.

Discussion

Hepatocellular adenocarcinoma is very common primary malignant tumor in adults which usually arises in liver after cirrhosis [1]. Though in pediatric population it is not that much common [4]. And multicentric hepatoma is very rare. In this population hepatoma commonly occurs during infancy but another peak is seen in early teen age but on background of underlying liver or systemic disease [9]. We have presented a case of an older child who does not have any underlying liver or systemic disease.

In children fibrolamellar carcinoma is more common which is considered as subtype of hepatocellular carcinoma [10]. In our case radiology as well as histopathology was very clear and did not favor fibrolamellar carcinoma rather showing features of typical hepatocellular carcinoma.

Regardless of the type of hepatocellular carcinoma, it is common in cirrhotic liver irrespective of the cause of cirrhosis [3]. In our case patient doesn't have underlying cirrhosis or any chronic liver disease. As patient doesn't had features of other metabolic or structural disease we didn't got workup for that like glycogen storage disease, Wilson disease and others. Also histopathology report was showing

underlying normal liver.

On triphasic CT abdomen hepatoma shows enhancement on arterial and washout on portovenous phase. While fibrolamellar carcinoma shows hypoattenuating mass with central scar and calcification [11].

Regarding treatment of hepatocellular carcinoma there are variable treatment modalities available including curative modalities like surgical resection, Radiofrequency ablation and liver transplantation [12]. Also for unresectable tumors locoregional therapies are available including percutaneous ethanol injection, transcatheter arterial chemoembolization and transcatheter arterial radioembolization [13].

The survival of patient after surgical resection depends upon size and number of tumor. As in our case tumor size was huge and it was multicentric almost involving 2/3rd of liver, this patient was at high risk for recurrence of disease and poor prognosis. The presence of cirrhosis is an independent poor prognostic factor for survival [14]. The favourable factors in this case are underlying liver was normal and clearance of tumor, as biopsy showed clear margins and no lymphovascular invasion of tumor. The literature has shown unsatisfactory long term prognosis because of tumor recurrence [15].

Conclusion

Though rare but Hepatocellular adenocarcinoma can occur in normal liver of young individuals. Early aggressive evaluation of lesion leads to diagnosis at curative stage. Although tumor was large but surgery should be considered as a favourable option if tumor is confined to liver.

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