Annals of Clinical Case Reports

Case Report
Published: 01 Mar, 2021

Sarcomatoid Hepatocellular Carcinoma: A Case Report and Imaging Findings

Parveen Kumar[1] and Nalini Bansal[2]*

[1]*Department of Radiodiagnosis and Imaging, Fortis Escort Heart Institute, India*

[2]*Department of Histopathology, Fortis Hospital, India*

Abstract

Sarcomatoid hepatocellular carcinoma is a rare malignancy with very poor prognosis. The study reports the case of a 54-year-old male who presented with post Trans Arterial Chemoembolization (TACE) residual/recurrent Hepatocellular Carcinoma (HCC). He had a history of hepatitis C for 10 years. Dynamic MRI showed two distinct lesions, one located in caudate lobe and another in segment VII. The lesion in caudate lobe showed delayed and prolonged peripheral enhancement with non-enhancing central component. The segment VII lesion showed faint arterial phase enhancement which persisted in portal venous phase without any washout. Patient was scheduled for liver transplantation. On histopathology the larger lesion showed spindle cells arranged in sheets having pleomorphic vesicular nuclei. Immunohistochemistry was positive for vimentin, AFP, Ck 8/18, Pan Ck, desmin and myogenin and thus a diagnosis of sarcomatoid hepatocellular carcinoma was established. Microscopy of segment VII lesion was consistent with high grade dysplastic nodule. This case describes the imaging features of sarcomatoid HCC and highlights the importance of histopathology and immunohistochemistry in final diagnosis.

Introduction

Sarcomatoid Hepatocellular Carcinoma is a rare neoplasm of liver comprising carcinomatous and sarcomatous components. It commonly develops in patients with background of alcoholic cirrhosis, chronic Hepatitis B Virus (HBV) and hepatitis C virus infection. A close relationship has been seen between the sarcomatous appearance in HCC and anticancer therapies. It has very poor prognosis due to high grade on histology and a higher rate of recurrence or metastasis. Therefore identifying this type is important for patient management and treatment planning. Herein, we present a case of sarcomatoid hepatocellular carcinoma. A detailed review of CT and MRI imaging characteristics with histopathological and immunohistochemical features are also discussed.

Case Presentation

A 54-year-old male patient was referred to our hospital for liver transplantation. He had a past medical history of hepatitis C which was detected in 2009 for which he was not taking any treatment. He subsequently developed abdominal distension and was found to have Hepatitis C Virus (HCV) related cirrhosis in 2017. He was kept on diuretics and antiviral therapy. He developed sustained virological response to HCV in 2019. He was doing well six months back when he developed loss of weight and weakness for which he was evaluated and found to have Hepatocellular Carcinoma (HCC) in segment V in outside hospital (Images not available). Alpha-Fetoprotein (AFP) level was elevated measuring 5368. According to the records he underwent Trans Arterial Chemoembolization (TACE) in august 2019 after which he developed sub capsular hematoma. He was managed conservatively for hematoma. A repeat CT was done there one month later which revealed background parenchymal cirrhotic changes, splenomegaly, splenic hilar and paraesophageal collaterals. A complex solid cystic lesion measuring 9.4 cm × 5.8 cm × 6.7 cm was noted in caudate lobe showing some soft tissue enhancement in delayed phases (Figure 1). Possibility of residual or recurrent tumor was suggested and he was referred to our hospital. On admission his general physical examination was normal. Vital parameters were unremarkable. Laboratory tests revealed elevated AFP (846; normal <10 ng/ml) and Protein induced by vitamin K absence-II PIVKA II (744.17; normal 17.36 mAU/ml to 50 mAU/ml). Biochemical indices of liver functions revealed mildly elevated SGOT (105; normal 15 IU/L to 37 IU/L), ALP (196; normal 50 IU/L to 136 IU/L) and GGT (162; normal 5 IU/L to 85 IU/L). Coagulation profile was deranged with elevated Prothrombin time (16; normal 11 sec to 13 sec) and fibrinogen (471; normal 200 mg/dl to 400 mg/dl). Antibody panel revealed non-reactive status for

OPEN ACCESS

Correspondence:
Parveen Kumar, Department of Radiodiagnosis and Imaging, Fortis Escort Heart Institute, New Delhi, India, Tel: +91-9971416939; E-mail: drparveenbatra@gmail.com

Received Date: *08 Feb 2021*
Accepted Date: *25 Feb 2021*
Published Date: *01 Mar 2021*

Citation:
Kumar P, Bansal N. Sarcomatoid Hepatocellular Carcinoma: A Case Report and Imaging Findings. Ann Clin Case Rep. 2021; 6: 1921.

ISSN: 2474-1655

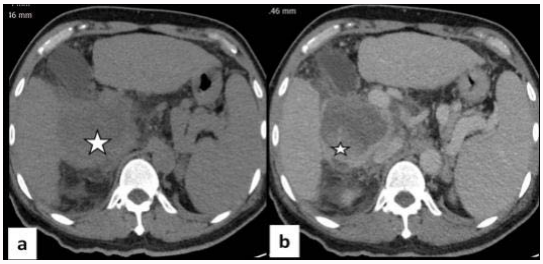

Figure 1: Computed Tomography abdomen findings: Non-Contrast Computed Tomography (NCCT) axial image (a) shows cirrhotic nodular liver dilated splenic vein and splenomegaly. A hypodense lesion is seen in caudate lobe (*). Contrast Enhanced Computed Tomography (CECT) image (b) at same level shows thick enhancing wall and eccentric enhancing component (*) in the lesion.

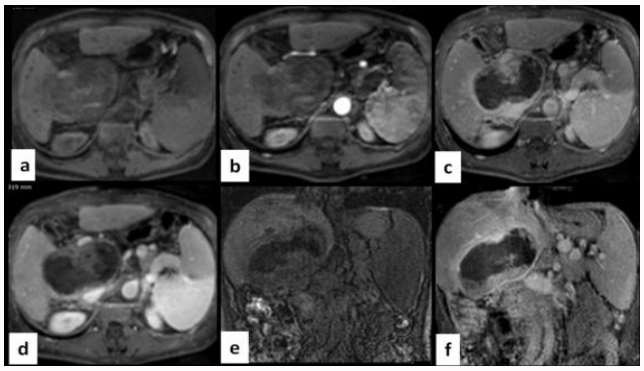

Figure 2: Dynamic MRI abdomen findings: A well defined lesion is seen in caudate lobe appearing: hetrointense on precontrastT1 (a), and showing progressive peripheral enhancement with eccentric enhancing soft tissue component on arterial (b), portal venous (c) and delayed phases (d). Same is shown in coronal pre-contrast T1 (e) and post contrast delayed phase T1 (f) images.

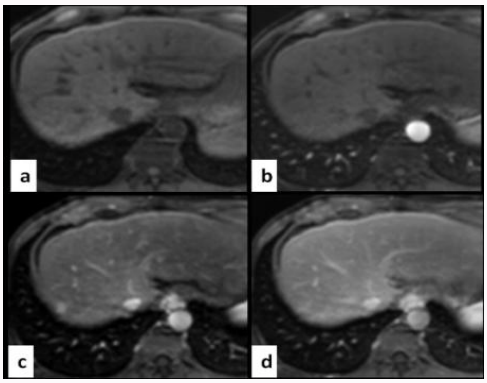

Figure 3: Dynamic MRI abdomen findings: Anotherill-defined lesion is seen in segment VII appearing iso-intense on pre-contrast T1 (a) and arterial phase (b). Homogenous enhancement is seen in portal venous phase (c) with no washout on delayed phase (d). Imaging findings are consistent with dysplastic nodule.

HbsAg, positive for HBcAb, reactive for HCV Ab and negative for HDV IgG. HBV DNA titer was <20 and titer for HCV RNA was <15. Rest of the biochemical investigations, ECG and echocardiography were unremarkable. A repeat contrast MRI was done with hepatobiliary specific contrast agent. The scan revealed background cirrhotic changes. Previously mentioned complex solid cystic lesion in caudate lobe appeared hetero-intense on T1/T2 with progressive delayed phase enhancement of peripheral solid component. Central

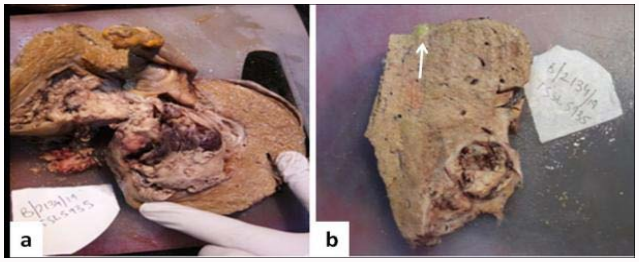

Figure 4: Gross findings (a) Gross showing large mass in a cirrhotic liver. (b) Segment VII nodule is also seen on liver surface (arrow).

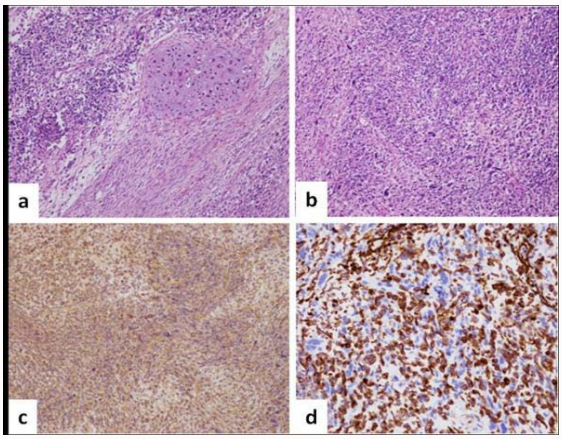

Figure 5: Microscopy findings: Microscopy showing areas of cartilaginous differentiation within the tumor (H&E × 20x) (a) Malignant spindle cell proliferation (H&E × 20x) (b), IHC AFP positivity (c) and IHC Vimentin positive in tumor cells (d).

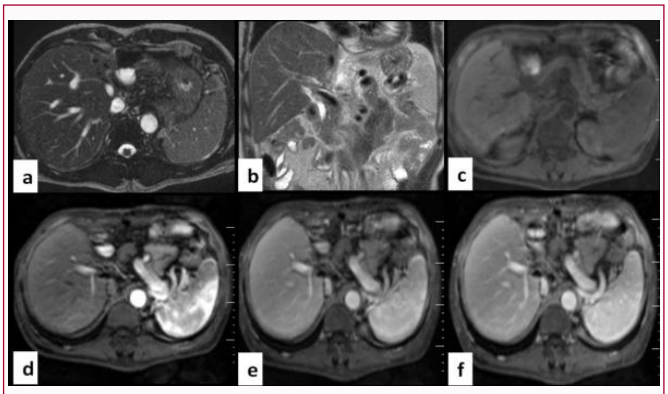

Figure 6: Dynamic MRI abdomen findings: Post liver transplantation images show normal appearance on axial T2 (a) and coronal T2 (b) images. No enhancing lesion is seen in liver on dynamic pre-contrast T1 (c), arterial (d), portal venous (e) and delayed phase (e).

part didn't enhance suggesting central necrosis (Figure 2). There was minimal increase in the size of lesion. Besides this one more lesions was seen in segment VII appearing isointense on T1/T2 with faint arterial phase enhancement which persisted in portal venous phase without any washout (Figure 3). The former lesion was characterized as residual HCC and later was characterized as dysplastic nodule. Liver transplantation was performed subsequently. The resected explant displayed firm consistency with brownish nodular external surface and irregular margins suggesting cirrhotic changes. A large variegated creamish yellow tumor mass showing hemorrhagic necrosis was noted in caudate lobe (Figure 4a). Another sub-

centimetric sized nodule was seen in segment VII in subcapsular location (Figure 4b). On microscopy the large tumor showed a neoplasm with spindle cells arranged in sheets having pleomorphic vesicular nuclei, prominent nucleoli and moderate cytoplasm (Figure 5a, 5b). On IHC the neoplastic spindle cells were positive for vimentin, AFP, Ck8/18 and focally positive for Pan Ck and arginase (Figure 5c, 5d). Desmin and myogenin were variably positive. Tumor cells were negative for S100, EMA, CD34 and HepPar-1. Overall the findings were suggestive for sarcomatoid hepatocellular carcinoma. Another nodule in segment VII showed features of high grade dysplastic nodule with preserved reticulin staining and negativity for Glypican 3. The patient was followed up and a repeat MRI was done one month after transplantation. No lesion was identified in transplanted liver and surgical bed (Figure 6). At present, the patient is in good condition.

Discussion

Hepatocellular Carcinoma (HCC) with spindle cell component has been referred to as sarcomatoid carcinoma. The pathogenesis of sarcomatoid carcinoma is not clear. There is a continuous debate on whether it is derived from the transition of an ordinary HCC to a sarcomatous morphology or it is a synchronous HCC and hepatic sarcoma. Presence of transitional features from HCC to spindle cell component and vimentin expression in the spindle cell on histopathology favors the dedifferentiation theory of sarcomatoid transformation in HCC [1,2].

It mainly occurs in elderly patients. The cumulative incidence is 0.79%. The risk factors include cirrhosis, chronic hepatitis B and chronic hepatitis C and are same like non sarcomatoid HCC [3]. KojiroM et al. [4] have shown close relationship between the sarcomatous appearance in HCC and anticancer therapies. The development of the sarcomatous appearance is presumed to be caused by the phenotypic change of HCC cells caused by anticancer therapy accelerating the proliferation of the sarcomatous cells existing in the original tumor as one of the histological components. The evidences suggesting this relationship are however poor in literature and there have been studies demonstrating its occurrence without any prior exposure to anti-cancer therapy [5].

Our patient also had history of hepatitis C infection for 10 years. He subsequently developed cirrhosis and HCC. Development of sarcomatoid HCC after TACE is in keeping with the relationship of TACE and sarcomatoid HCC described in literature.

Sarcomatoid HCC is known to exhibit central necrosis and hemorrhage owing to its rapid growth and poorly differentiated morphology. Imaging findings on CT and MRI are in keeping with this growth pattern. The typical imaging features on CT include peripheral ring-like enhancement and central non-enhancing region. The peripheral enhancing tissue represents viable neoplastic tissue with fibrous stroma, and internal unenhanced region correspond to coagulation necrosis and hemorrhage. The peripheral enhancing tissue shows delayed and prolonged enhancement which correlates with extensive interstitial space allowing slow diffusion of contrast between vascular and interstitial spaces. The type of enhancement pattern is also seen in hemangiomas, metastases and in intrahepatic cholangiocarcinoma however, the delayed peripheral enhancement is more commonly seen in sarcomatoid HCC [6,7].

On MRI, sarcomatoid carcinoma shows lobulated or irregular shapes. It shares few imaging features with intrahepatic cholangiocarcinoma like lobulated shape, rim-like arterial-phase hyper enhancement, large necrotic area (if the lesion is large), delayed central enhancement (if the lesion is small), biliary dilatation and intrahepatic and extrahepatic metastases, although targetoid appearance or capsular retraction are less common than in intrahepatic cholangiocarcinoma [8].

In present case the large lesion in caudate lobe showed typical imaging features of sarcomatoid HCC. Progressive peripheral enhancement was seen on both CT and MRI images. Central part didn't enhance suggesting central necrosis.

On histopathology the sarcomatoid HCC demonstrates transitional features between sarcomatoid tumor and ordinary HCC. Immunohistochemical study for vimentin, cytokeratin, alpha fetoprotein and epithelial membrane antigen are helpful in the pathological diagnosis. It has been seen that sarcomatoid lesions are positive for both cytokeratin and vimentin which are the markers of epithelial and mesenchymal tissue respectively [5]. The tumor in the present case was diffusely positive for vimentin and alpha-fetoprotein, which is consistent with sarcomatoid HCC.

Similar histopathological findings were seen in present case. On microscopy the large tumor showed a neoplasm with spindle cell arranged in sheets having pleomorphic vesicular nuclei, prominent nucleoli and moderate cytoplasm. On IHC the neoplastic spindle cells were positive for vimentin, AFP, Ck8/18, Pan Ck, desmin and myogenin.

It has very poor prognosis. There are higher chances of lymph node and distant metastasis. There are high chances of early recurrence even after curative therapy, such as RFA, surgical resection, or liver transplantation. Study done by Liao et al. [3] estimated that 1, 3, and 5 year recurrence-free survival rates of sarcomatoid HCC were 50.0%, 15.0%, and 5.0% as compared to 83.0%, 44.3%, and 22.6% for the non-sarcomatoid group. The clinical importance of this diagnosis lies in the fact that it is an independent predictor of recurrence after curative treatment and all-cause mortality.

There was no recurrence seen in follow up MR imaging done after 1month of post transplantation. He is advised another follow up scan after 6 months.

In conclusion, sarcomatoid HCC is an aggressive tumor with high recurrence rate and tendency for rapid growth. It should be suspected whenever there is history of TACE therapy. The typical imaging features should prompt the diagnosis although the final word is from tissue sampling and intensive immunohistochemical analysis.

Learning Points

1. The risk factors for sarcomatoid HCC include cirrhosis, chronic hepatitis B and chronic hepatitis C and are same like non sarcomatoid HCC.

2. The typical imaging features on CT include delayed and prolonged peripheral ring-like enhancement and central non-enhancing region.

3. On histopathology the sarcomatoid HCC demonstrates transitional features between sarcomatoid tumor and ordinary HCC. Generally, components of sarcomatoid lesions are positive for both cytokeratin (epithelial marker) and vimentin (mesenchymal marker).

4. It has poor prognosis with higher chances of lymph node and distant metastasis.

References

1. Maeda T, Adachi E, Kajiyama K, Takenaka K, Sugimachi K, Tsuneyoshi M. Spindle cell hepatocellular carcinoma. A clinicopathologic and immunohistochemical analysis of 15 cases. Cancer. 1996;77(1):51-7.
2. Kakizoe S, Kojiro M, Nakashima T. Hepatocellular carcinoma with sarcomatous change. Clinicopathologic and immunohistochemical studies of 14 autopsy cases. Cancer. 1987;59(2):310-6.
3. Liao SH, Su TH, Jeng YM, Liang PC, Chen DS, Chen CH, et al. Clinical manifestations and outcomes of patients with sarcomatoid hepatocellular carcinoma. Hepatology. 2019;69(1):209-21.
4. Kojiro M, Sugihara S, Kakizoe S, Nakashima O, Kiyomatsu K. Hepatocellular carcinoma with sarcomatous change: A special reference to the relationship with anticancer therapy. Cancer Chemother Pharmacol. 1989;23:S4-8.
5. Yokomizo J, Cho A, Yamamoto H, Nagata M, Takiguchi N, Kainuma O, et al. Sarcomatous hepatocellular carcinoma without previous anticancer therapy. J Hepatobiliary Pancreat Surg. 2007;14(3):324-7.
6. Honda H, Hayashi T, Yoshida K, Takenaka K, Kaneko K, Fukuya T, et al. Hepatocellular carcinoma with sarcomatous change: Characteristic findings of two-phased incremental CT. Abdom Imaging. 1996;21(1):37-40.
7. Koo HR, Park MS, Kim MJ, Lim JS, Yu JS, Jin H, et al. Radiological and clinical features of sarcomatoid hepatocellular carcinoma in 11 Cases. J Comput Assist Tomogr. 2008;32(5):745-9.
8. Seo N, Kim MJ, Rhee H. Hepatic sarcomatoid carcinoma: magnetic resonance imaging evaluation by using the liver imaging reporting and data system. Eur Radiol. 2019;29(7):3761-71.