

# Carcinosarcoma of Liver – A Rare Entity

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## ABSTRACT

Liver carcinosarcoma is a rare entity, with only 42 cases reported so far. World Health Organization (WHO) has defined carcinosarcoma as a mixture of both carcinomatous and sarcomatous components. They have an aggressive behaviour with poor outcome. Unlike hepatocellular carcinoma, there is no characteristic imaging feature to diagnose hepatic carcinosarcomas. Surgery remains the only curative option. There is no established optimal chemotherapy regimen for carcinosarcoma. Here we present two cases of primary hepatic carcinosarcoma with extensive metastases.

**Keywords:** Chemotherapy role, Hepaticcarcinosarcoma, Pathological features, Poor prognosis, Radiological features.

*Asian Pac. J. Health Sci.*, (2020); DOI: 10.21276/apjhs.2020.7.2.6

## INTRODUCTION

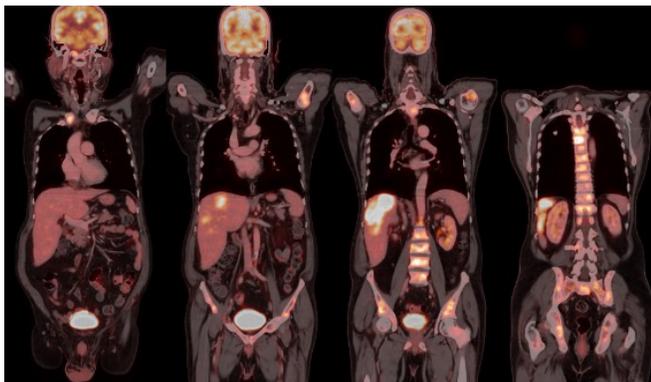
Liver carcinosarcomas were first described by Craig *et al.* in 1989. He defined carcinosarcoma of the liver as a mixture of hepatocellular carcinoma (HCC) and non-spindle cell sarcoma components with specialized differentiation into malignant muscle, cartilage or bone component. WHO defines human chorionic somatomammotropin (HCS) as a mixture of carcinomatous components, either HCC or comprehensive care center (CCC) and sarcomatous components.

Hepatic carcinosarcoma is a rare malignancy with only 42 case reports so far. HCS is more common in men than women with an incidence ratio of 3:1. 50% were positive for Hepatitis B and 40% exhibited elevated AFP levels.<sup>[1]</sup> Nearly 70% of patients with HCS had advanced disease at presentation. The most common sites of metastases were lung and lymph nodes.

We present two case reports of metastatic hepatic carcinosarcoma.

## CASE REPORT-1

A 66-year-old male patient, known diabetic and hypertensive, presented with the complaints of low backache for a month and was evaluated. Blood investigations showed hypoalbuminemia and leukocytosis. Ultrasound abdomen showed multiple hypoechoic nodular lesions in the liver. Magnetic resonance imaging (MRI) spine was suggestive of multiple bone secondaries. He was further evaluated with Positron emission tomography-computed tomography (PET-CT) whole body, which showed an intrahepatic mass probably cholangiocarcinoma with liver and bone secondaries. CA19-9 was mildly elevated at 55.8U/mL. Biopsy and IHC from the liver mass were consistent with Carcinosarcoma (Pancycytokeratin AE1 + AE3 – positive, CK7 positive, vimentin positive, FL-1 positive,



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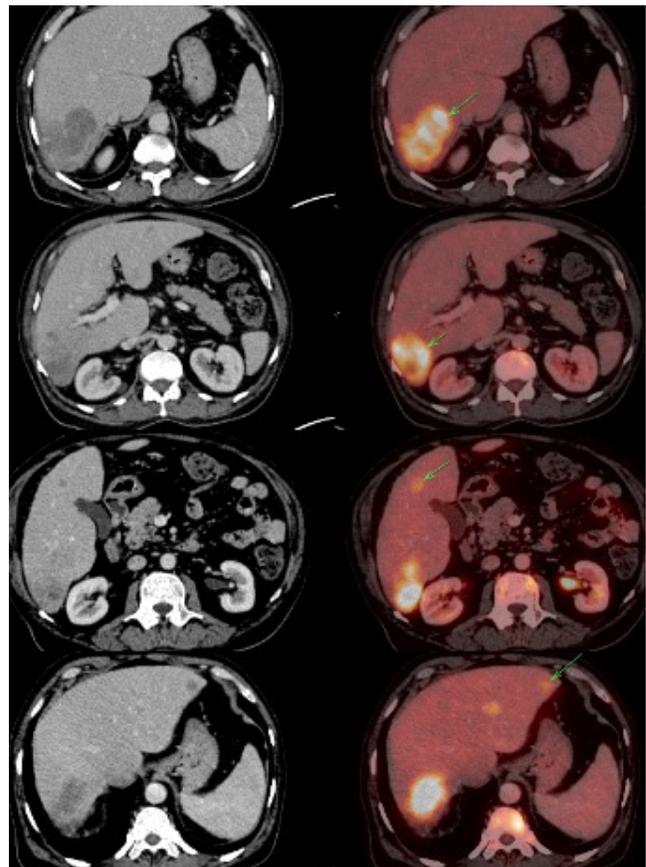
**How to cite this article:** Ramya A, Roopa BMS, Yabesh TA, Raja T. Carcinosarcoma of Liver – A Rare Entity. *Asian Pac. J. Health Sci.*, 2020; 7(2):26-28

**Source of support:** Nil

**Conflict of interest:** None

**Received:** 15/03/2020 / **Revised:** 20/04/2020 **Accepted:** 28/04/2020

SMA positive). He was started on palliative chemotherapy with gemcitabine and cisplatin (Figures 1 and 2).



**Figure 1 and 2:** Positron emission tomography-computed tomography suggestive of intrahepatic mass probably cholangiocarcinoma with liver and bone secondaries.

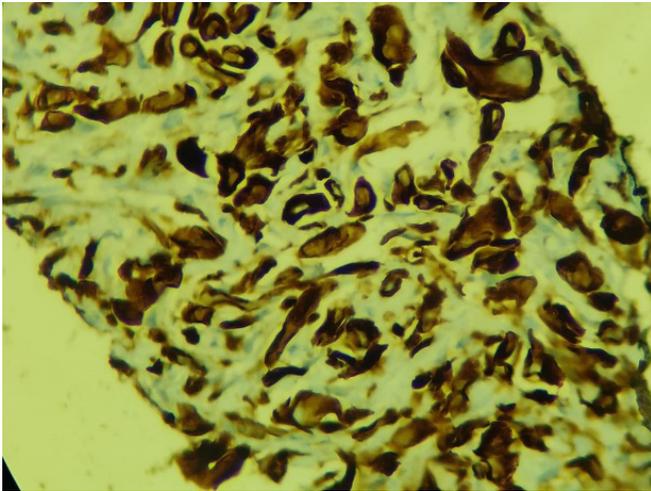


Figure 3: IHC for PAN CYTOKERATIN

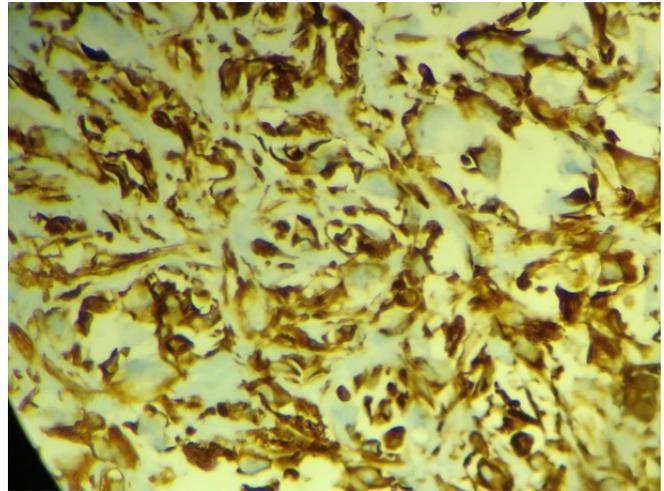


Figure 4: IHC for VIMENTIN

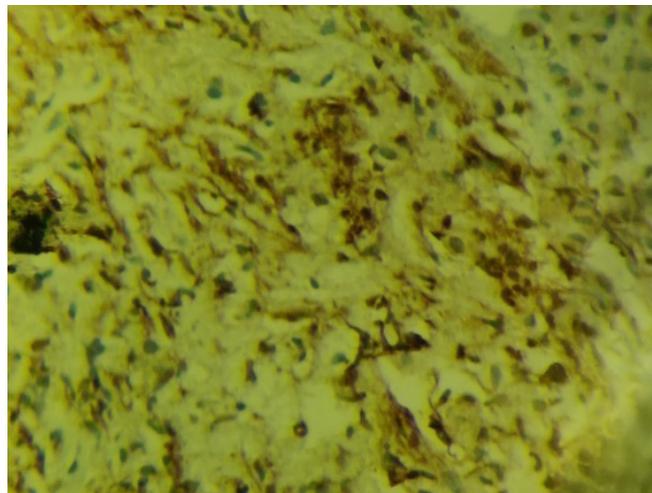


Figure 5: IHC for SMA

## CASE REPORT 2

A 57-year-old male had presented with complaints of loss of weight and appetite for a month and was evaluated. Blood investigations showed direct hyperbilirubinemia. Ultrasonography (USG) abdomen showed multiple hypoechoic liver lesions with IHBR dilatation and splenomegaly. CT abdomen showed multiple hypodense lesions in both lobes of liver with IBR dilatation and diffuse GB wall thickening. PET CT raised the possibility of hilar cholangiocarcinoma with hepatic adrenal and skeletal mets. Carcinoembryonic antigen (CEA) was elevated at 22.6 ng/mL. Biopsy and IHC was suggestive of carcinosarcoma (Figure 3-5).

He was initially managed with ERCP and stenting and was started on palliative radiotherapy for bone lesions. He was planned for palliative chemotherapy after recovery of liver function but deteriorated further with hepatic encephalopathy.

## DISCUSSION

Hepatic carcinosarcoma is an aggressive disease with a mean age of onset between 33–75 years<sup>[1]</sup> and is more common in males. Carcinosarcoma is more common in the uterus, ovary, and urinary bladder.<sup>[2]</sup>

The pathogenesis of HCS is controversial. Majority develop in non-cirrhotic liver. This findings suggest that HCS develops from a stem cell or multipotent hepatic progenitor cell. The histopathology shows both carcinomatous and sarcomatous components with HCC being the most common carcinomatous component. In general, HCS is an aggressive disease and has a poor prognosis. CT is the most common imaging modality used. HCS is a heterogeneous tumor with cystic, solid, and necrotic areas. Calcification and osteogenesis were observed in 50% of cases especially in those with osteosarcomatous components. Unlike HCC, HCS rarely exhibits a capsule, which might indicate its aggressive behavior.<sup>[1]</sup>

The proposed histogenetic mechanisms involved in the co-existence of carcinoma and sarcoma components in the same tumor include 'transformation', 'combination' and 'collision'.<sup>[3]</sup> 'Transformation' means that, part of the carcinoma undergoes sarcomatous transformation. 'Combination' signifies that a tumor arises from a single stem cell that differentiates into epithelial and mesenchymal tissues. 'Collision' denotes that carcinoma and sarcoma elements of distinct origin invade each other.<sup>[3]</sup>

There are no characteristic imaging features for HCS. A large tumor with more extensive cystic and necrotic degeneration, without capsule, with hypervascular enhancement, more frequently

presented with lymphadenopathy and invasion of adjacent organs, these findings may be helpful to distinguish HCS from other malignancy.<sup>[1]</sup>

Yasutake *et al.* reported a case of liver carcinosarcoma including an HCC-component, which was hyperintense in the hepatobiliary-phase on gadolinium-ethoxy benzyl diethylene triaminepentaacetic acid (Gd-EOB-DTPA) contrasted enhanced MRI. Further studies are required to establish the role of Gd-EOB-DTPA enhanced MRI in distinguishing HCS from HCC.<sup>[1]</sup>

CT presentation of HCS is nonspecific. Sumiyoshi *et al.* observed that carcinosarcoma on magnetic resonance imaging revealed hypointensity on T1-weighted images and heterogeneous hyperintensity on T2-weighted images.<sup>[4]</sup> Shu *et al.* observed that in one case of liver carcinosarcoma, triple-phase contrast CT revealed a mixed density mass with inhomogeneous enhancement, multiple cystic nodules and irregular rim enhancement.<sup>[5]</sup>

Primary curative resection is the only curative option for HCS. Lymphadenopathy is more common with carcinosarcoma, and lymphadenectomy improves prognosis in localized hepatic carcinosarcoma.<sup>[2]</sup> In those with the limited disease who undergo surgery, the meantime to recurrence was 4.9 months (range: 1 – 15 months).<sup>[2]</sup> In one Japanese case report, the recurrences were nodal, and repeated surgical excisions managed them. All the nodal disease had only sarcomatous elements. In view of the aggressive behavior with the dissemination of the sarcomatous elements, apart from surgery systemic therapy also needs to be considered. The optimal chemotherapy program for HCS is still under debate.

Hepatic carcinosarcoma can undergo dedifferentiation at the site of metastases into sarcoma.<sup>[6]</sup>

Here, both the patients presented with advanced disease and one showed rapid deterioration from the time of symptom onset, giving us very little time to intervene.

## CONCLUSION

Hepatic carcinosarcoma is a rare, aggressive disease with poor outcomes. Carcinosarcoma is a histopathological diagnosis. Further studies are required to identify characteristic image findings to diagnose carcinosarcoma and to understand the most suitable chemotherapy regimen that will produce an adequate response and survival benefit.

## REFERENCES

1. Li J, Liang P, Zhang D, Liu J, Zhang H, Qu J, *et al.* Primary carcinosarcoma of the liver: imaging features and clinical findings in six cases and a review of the literature. *Cancer Imaging*. 2018 Feb 27;18(1):7.
2. Kurita D, Mokuno Y, Matsubara H, Kaneko H, Shamoto M, Satou A, *et al.* Primary hepatic carcinosarcoma with multimodal treatment. *Nagoya J Med Sci*. 2018 Aug;80(3):423-429.
3. She R, Szakacs J. Carcinosarcoma of the liver: a case report and review of the literature. *Arch Pathol Lab Med* 2005;129:790 –793.
4. Sumiyoshi S, Kikuyama M, Matsubayashi Y, *et al.* carcinosarcoma of the liver with mesenchymal differentiation. *World J Gastroenterol*. 2007;13(5):809–812. DOI:10.3748/wjg.v13.i5.809
5. Liu LP, Yu XL, Liang P, Dong BW. Characterization of primary hepatic carcinosarcoma by contrast-enhanced ultrasonography: A case report. *World J Gastroenterol* 2014; 20(6): 1630-1634
6. Kwon JH, Kang YN, Kang KJ. Carcinosarcoma of the liver: a case report. *Korean J Radiol*. 2007;8(4):343–347. DOI:10.3348/kjr.2007.8.4.343