A Rare Case of Fibrolamellar Hepatocellular Carcinoma with Unusual Presentation in a Young Indian Female- a case report

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Abstract

Fibrolamellar carcinoma (FLC) is a rare subtype of Hepatocellular carcinoma (HCC) comprising Approx 1% of HCC. This variant of HCC commonly metastasizes to regional lymph nodes (celiac, gastric, para aortic). But metastasis to cervical lymph nodes is very rare. It is also very rare in Asian population. We are reporting a case of a rare cervical lymph node metastasis from FLC in an Indian female. Patient was a 20 yrs female presented with a cervical lymph node enlargement. USG and CT revealed lesion in liver and massive lymphadenopathy involving regional and para aortic group of lymph nodes. Diagnosis was made on histopathological examination of cervical lymph node biopsy.

Key words: Fibrolamellar carcinoma, hepatocellular carcinoma, Hepatic carcinoma

Introduction

Fibrolamellar carcinoma (FLC) is a rare variant of HCC comprising Approx 1% of HCC [1]. It is common in US and Europe [1]. In Asia it is very rare. It was initially described by Edmondson in 1956 [2]. Clinically FLC usually present with abdominal pain and metastasize to regional lymph nodes (Celiac, gastric, paraaortic).

Metastasis to cervical lymph node is rarely observed in FLC. The most characteristic Histologic feature of FLC is fibrosis arranged in a lamellar fashion around the neoplastic hepatocytes.

The tumor cells are large polygonal with abundant eosinophilic cytoplasm, vesicular nuclei and prominent nucleoli [3]. We have reported a rare variant of HCC (i.e. FLC) in an Indian female presented with cervical lymph node metastasis which is an unusual presentation.

Case report

We are reporting a case of 20 yrs Indian female presented with a lump in the upper cervical region. There were no other complaints except for mild abdominal discomfort. Systemic evaluation revealed hepatomegaly. Her laboratory test showed CBC and ESR within normal limits. Liver profile and kidney profile were also normal.

USG report showed pre and para-aortic lymphadenopathy with borderline hepatomegaly and septate gallbladder filled with sludge. USG guided FNAC was done which showed plenty of normal hepatocytes along with acute and chronic inflammatory cells in a necrotic background. Smear was Negative for malignancy.

CT report showed multiple nodules in the liver. There were significant lymph nodes along the portal vein that were seen to compress the common bile duct with mild dilatation of intrahepatic bile ducts. Gallbladder was dilated and its fundus showed significantly thicken wall.

Massive lymphadenopathies was seen within retroperitoneum from renal vein level to the level of aortic bifurcation. Lymphnodes were also seen along IVC and coelic plane & peripancreatic area.

There was ascites and bilateral pleural effusion. CT guided FNAC from left paraaortic nodes was done but it was not informative as it showed only haemorrhagic aspirate. After seeing the reports clinicians were suspecting it to be a case of Hodgkins lymphoma and excision biopsy from enlarged cervical lymph node was send to our department for histopathological examination which revealed metastatic deposits from Fibrolamellar variant of hepatocellular carcinoma.
Histopathology Report

H/E stained sections from cervical lymph node biopsy show malignant epithelial cell effacing the normal lymph node architecture. Cells are large polygonal having large vesicular nuclei with prominent nucleoli and abundant eosinophilic cytoplasm. The cells are arranged in trabecular and acinar pattern, and are separated by fibroconnective tissue. Features are suggestive of metastatic deposits from Fibrolamellar variant of Hepatocellular carcinoma.

Discussion

FLC is a rare morphologic variant of HCC which is not associated with cirrhosis or other chronic liver disease and having a generally favorable prognosis [3, 4, 5, 6]. It occurs commonly in children and young adult between 5-35yrs and has equal sex incidence [3]. However some studies have shown slight female predominance [1, 4, 7]. Etiology and the genetic abnormality of FLC are unknown. Studies have shown that they lack mutation in the gene most commonly mutated in typical HCC (TP53 & CTNN B 4). FLC generally present with a nonspecific clinical signs and symptoms, which include abdominal pain, fatigue, malaise and weight loss. Overall the most common physical finding is an abdominal mass or hepatomegaly. However a wide variety of unusual presentations have also been described.

Laboratory findings in FLC are usually normal. In our case also liver profile was normal. Alpha feto protein (AFP) levels are typically normal in FLC [8]. Craig et al highlighted the distinctive histologic features of FLC.
which consist of deeply eosinophilic neoplastic hepatocytes and fibrosis arranged in a lamellar fashion around the neoplastic hepatocytes [3]. Because of its characteristic histological features most case of FLC are readily diagnosed from metastatic site. Cytologically FLC is characterized by oncocytic hepatocytes which are three times the size of normal hepatocytes [9, 10].

The treatment of choice in hepatic lesion is complete resection. Inoperable cases of FLC are benefited from adjuvant chemotherapy. FLC has high operability rate and better prognosis compare to conventional HCC [3, 4, 11].

Kakar S et al in his study found that prognosis of FLC and conventional HCC without cirrhosis were similar which reflect that probably the better prognosis of FLC is due to lack of cirrhosis in it [12].

Our case of FLC presented with cervical lymph node metastasis which is a rare presentation. To our knowledge very few cases of FLC has been reported from India. One largest series of 6 cases was reported by Singhal et al in 2002 [13].

Two more cases were reported, one was FLC associated with Non Bacterial Thrombotic Endocarditis in a young 17yr male [14] and the other case was of a 25 years Indian female in which diagnosis was made based on cytological features of FLC [10].

Conclusion

Clinical recognition of this variant of HCC is important because of excellent result of complete surgical resection. And a definite diagnosis of this variant of HCC can be made only by histopathological examination of the lesion. Clinicians should keep this variant of HCC in mind in young adult without underlying hepatitis or cirrhosis.

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