Carcinoma Ovary Masquerading as Metastatic Hepatocellular Carcinoma—Case Report

ABSTRACT

Malignant tumours with hepatoid features resembling hepatocellular carcinoma (HCC) have been described in different sites. These tumours are very rare and are found particularly in the stomach, ovary, pancreas, renal pelvis and urinary bladder. Hepatoid variant of carcinoma ovary was first of all reported by Isikura and Scully in 1987. Till date, with the best of our knowledge, only 21 patients with diagnosis of Hepatoid carcinoma of the ovary have been reported. No case has been reported from India till date. This is a case of 56 years female presented to us with a complaint of abdominal distension, pain and weight loss of 2 months duration. Omental Biopsy was suggestive of HCC. On IHC, tumour cells were positive for HepPar 1, Glypican 3 and focal positivity for CK7 while negative for SALL4, PAX8, CK20 and TTF 1. She underwent interval cytoreductive surgery in January 2014. The final HPE report was hepatoid variant of carcinoma ovary.

KEYWORDS: hepatoid carcinoma ovary

INTRODUCTION

Malignant tumours with hepatoid features resembling hepatocellular carcinoma (HCC) have been described in different sites. These tumours are very rare and are found particularly in the stomach, ovary, pancreas, renal pelvis and urinary bladder. Hepatoid variant of carcinoma ovary was first of all reported by Isikura and Scully in 1987, described a special type of ovarian carcinoma that they called hepatoid based on two criteria: tumour cells with abundant eosinophilic cytoplasm that resembled HCC and positive staining for AFP by immunohistochemistry. Till date, with the best of our knowledge, only 21 patients with diagnosis of hepatoid carcinoma of ovary have been reported. No case has been reported from India till date.

CASE REPORT

56-year-old female without any co-morbidities presented to us with a complaint of abdominal distension, pain and weight loss of 2 months duration. She was investigated outside with CECT abdomen, which revealed peritoneal nodules with mesenteric thickening, ascites and normal architecture of the liver. She was investigated at our hospital with serum AFP which was 6,132 ng/ml, serum CEA, which was 3 ng/ml, serum CA 125, which was 5,613 U/ml, serum CA 125 was 54.3 U/ml and serum AFP was 4323 ng/ml. She underwent interval cytoreductive surgery in January 2014. The final HPE report was hepatoid variant of carcinoma ovary.
Hepatoid carcinoma of the ovary is a very rare type of high-grade, invasive malignant ovarian tumour composed mainly of epithelioid cells. This entity, based on morphology and immunohistochemistry is similar to HCC, can be associated with high levels of AFP and carries a poor prognosis. A focal hepatoid differentiation is relatively common in yolk sac tumours, but infrequent in other germ cells and epithelial ovarian tumours.

It is a very rare ovarian tumour that usually presents with signs and symptoms of an adnexal mass, such as progressive abdominal distension and lower abdominal pain and has been described in post-menopausal women aged from 42 to 78 years with an average age of 63 years. It can also be detected as an asymptomatic unilateral ovarian mass (rarely bilaterally) with the elevated serum level of α-fetoprotein. This malignancy mostly was found in an advanced clinical stage and progresses rapidly, with metastases to the abdomen and occasionally to the lungs; with median survival about 2 years.

Grossly, the tumour may appear as entirely solid or with cystic areas, and there may be multiple foci of haemorrhage and necrosis. Microscopically, these tumours are characterized by solid sheets of large cells with abundant eosinophilic cytoplasm, centrally pleomorphic nuclei and distinct cellular borders. On immunostaining, the tumour is focally positive for AFP and polyclonal CEA (pCEA) and is cytokeratin positive. Hepatocyte paraffin-1 (HepPar 1) is positive in primary ovarian carcinoma with hepatoid differentiation, as well as HCC, and so this marker is not useful to distinguish metastatic HCC from primary hepatoid variant of carcinoma ovary. A diffusely positive staining pattern for CK7 is highly consistent with hepatoid variant of carcinoma ovary.

The differential diagnoses of hepatoid variant of carcinoma ovary include hepatoid yolk sac tumour (HYST), clear-cell carcinoma, lipid cell tumour, endometrial carcinoma and undifferentiated carcinoma. HYST is another AFP-producing tumour with hepatoid differentiation, which is very similar to hepatoid variant of carcinoma ovary. HYST stains negative for HepPar 1 on IHC, which is a major differentiating point between HYST and hepatoid variant of carcinoma ovary. The occurrence of the HYST in younger patients, some with gonadal dysgenesis and the presence of a residual component of typical yolk sac tumour or a polyvesicular vitelline pattern, or glandular-like structure some with mucin production, is helpful for differential diagnosis. In contrast, hepatoid variant of carcinoma ovary occurs generally in older or post-menopausal woman and is not associated with germ cell tumours or gonadal dysgenesis. It is difficult to distinguish metastatic HCC from hepatoid variant of carcinoma ovary. HCC rarely metastasizes to the ovary and elevation of CA-125, a marker for ovarian surface epithelial tumours would support an ovarian origin. Usually, tumour cells of hepatoid variant of carcinoma ovary are positive for AFP staining, and the presence of focal staining for AFP was considered to be essential when diagnosing AFP. However, not all hepatoid variant of carcinoma ovary are associated with AFP positive, AFP is negative in some cases. Undifferentiated carcinoma
with abundant eosinophilic cytoplasm, which is negative for AFP staining, is also considered as hepatoid carcinoma. To date, there is insufficient data regarding the optimal treatment of patients with hepatoid variant of carcinoma ovary. Most patients are treated by surgery, followed by a chemotherapy regimen similar to those used in patients with ovarian-like carcinomas. For these patients, the initial step in patient management is reduction of the surgical bulk, followed by chemotherapy. Sorafenib has been tried in a few patients, but response rate has been very low. In our case also, there was a minimal response to targeted agent Sorafenib with very short progression free interval of 2 months. This suggests that despite a pathologic resemblance between hepatoid variant of carcinoma ovary and HCC, they are biologically different and that hepatoid variant of carcinoma ovary clinically behaves more like an epithelial ovarian tumour than a hepatocellular tumour and should be treated as such. Additional efficacy may be provided by intraperitoneal chemotherapy through the combination of new cytotoxic agents with paclitaxel and carboplatin, and integrating biological agents in front-line therapy.

REFERENCES