Primary hepatic choriocarcinoma: a rare cause of spontaneous haemoperitoneum in an adult

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Abstract
Choriocarcinoma is a beta human chorionic gonadotrophin secreting neoplasm pertinent to uterus and pregnancy mostly. It occurs primarily in gonads but rarely in extragonadal sites. Primary hepatic choriocarcinoma is an extremely rare tumor. Most of the reported cases are seen in infants representing metastasis from an occult placental choriocarcinoma. Till date, only 7 cases of primary hepatic choriocarcinoma in adults have been reported in literature. We present a case of a 40-year-old male presenting as haemoperitoneum due to ruptured hepatic tumor. He underwent emergency left lateral segmentectomy. He died on 10th postoperative day. The surgical specimen and autopsy findings confirmed it to be primary hepatic choriocarcinoma. This is the first case report from Indian Subcontinent. A brief case report and review of literature is presented.

Introduction
Choriocarcinoma, primarily occurs in gonads but infrequently may occur in extragonadal sites. The sites include mediastinum, retroperitoneum or pineal gland and rarely in parenchymal organs. Initially, they were considered as metastasis from an occult primary tumor in gonads, but now it is known that they arise either as a consequence of abnormal migration of germ cells during embryogenesis or from a different histogenetic origin.

Choriocarcinoma of liver without a detectable primary tumor in gonads, retroperitoneum or mediastinum is termed as primary hepatic choriocarcinoma. This is a rare tumor seen more often in infantile age group. Few cases in adults have been reported. Due to varied clinical presentations and extreme rarity of the tumor, diagnosis on small needle biopsy is difficult. We present a case of adult male diagnosed to have primary hepatic choriocarcinoma on histopathology and immunohistochemical studies.

Case Report
A 40-year-old male was referred to us with history of sudden onset pain in abdomen. He was admitted in private hospital where computed tomography (CT) scan of abdomen was done which was suggestive of ruptured hepatic choriocarcinoma in left lobe of liver (Figure 1). Patient was resuscitated and given adequate blood transfusions and was referred to our centre. On admission his vitals were stable, but pallor was present. Abdomen was distended with generalized tenderness and rigidity. He was posted for an emergency surgery. Abdominal exploration revealed about 1.5 to 2 L of haemoperitoneum and multiple nodular lesions in both lobes of liver with large ruptured lesions in left lobe of liver with active bleeding. Rest of the abdomen was normal. After achieving adequate inflow and outflow control, left lateral segmentectomy was done in view of active bleeding (Figure 2). Patient withstood procedure well. Histopathology revealed atypical trophoblastic cells predominantly cytotrophoblast and few syncitiotrophoblast lying in sheets as well as in clusters with large number of mitotic figures along with the normal hepatocytes suggestive of high grade malignant tumor (Figure 3). Immunohistochemistry was positive for beta human chorionic gonadotrophin (Figure 4) and negative for CK, EMA, CD-30, AFP, CD-31 and CD-34 suggestive of choriocarcinoma. His serum HCG levels were significantly raised however serum levels of AFP, CEA and CA 19-9 were normal. Patient died on postoperative day 10 due to sudden cardiopulmonary arrest. On autopsy, multiple nodular lesions were present on the remaining liver. Rest of the abdominal organs, retroperitoneum and mediastinum were unremarkable. Bilateral testes were grossly normal which on subsequent serial sectioning and histological examination did not show any evidence of pathologic features associated with germ cell tumor regression or a scar. Based on histopathological report and autopsy findings it was diagnosed as primary hepatic choriocarcinoma.

Discussion
Primary hepatic choriocarcinoma is an extremely rare tumor. Majority of them may represent metastasis from an occult placental choriocarcinoma. Primary hepatic choriocarcinoma in adults are known to arise from abnormal migration of germ cells during embryogenesis or different histogenetic origin.

Clinically, patient may present with right upper abdominal pain and/or abdominal lump or distension. Those with advanced disease may present with symptoms due to metastasis to various organs like brain and lungs. Infertility, gynaecomastia and features of thyrotoxicosis may present in some patients, attributed to over production of HCG by tumour cells. Very rarely, it may rupture spontaneously producing haemoperitoneum and may present as an acute abdomen, as in our case.

Major issue in diagnosing primary hepatic choriocarcinoma is to exclude metastasis from an occult primary in gonads by serial sectioning and histological examination, as these tumors may be small or undergo spontaneous regression at the time of metastasis.

Diagnosis on small needle biopsy is difficult due to rarity of the tumor in liver. Immunohistochemical staining for beta HCG, PLAP, HPL, Hepar-1, may aid the diagnosis and helps to differentiate from the other tumours which mimic choriocarcinoma like giant cell variant of poorly differentiatated hepatocellular carcinoma, or tumor with trophoblast like giant cells. Finally imaging modalities like ultrasonography and CT scan may be useful to assess the extent of primary tumour and metastasis to various organs.

Treatment consists of complete surgical resection of tumor if localized to liver and without ascites followed by chemotherapy. For
advanced or metastatic disease chemotherapy is given. Chemotherapeutic drugs commonly used are etoposide, methotrexate, actinomycin-D, cyclophosphamide.

Prognosis of primary hepatic choriocarcinoma is distinctly poor as compared to its testicular counterpart owing to lack of restrictive effect of tunica albugenia as in the testes; hence they attain a large size and often invade adjacent vital structures by the time diagnosis is made. Average survival being 2 to 8 months as reported in literature, hence further studies are needed for early diagnosis and better treatment to improve survival.

References