Ruptured primary hepatic yolk sac tumor, a case report of unusual presentation and 3 years follow up

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Abstract: Endodermal sinus tumor (EST), also known as yolk sac tumor (YST), is a member of the germ cell tumor group of cancers. Primary yolk sac tumour of the liver is extremely rare, and when it occurs in a young child it can be confused with hepatoblastoma. A 27-year-old woman presented with a liver mass and histological examination of the tumor revealed the morphological and immunohistochemical features of a yolk sac tumour. There was no evidence of an extrahepatic primary source. Neoadjuvent chemotherapy was conducted and upon first cycle tumor get ruptured and underwent debulking. Three line of chemotherapy used to achieve radiological and laboratory control.

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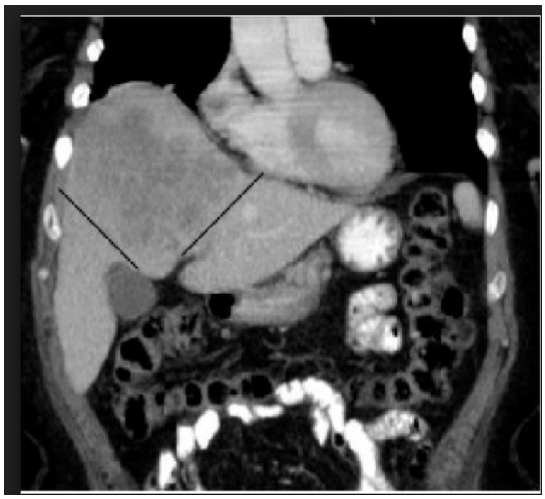
Case Report

A 27-year-old lady was otherwise healthy. started to notice Rt upper Quadrant swelling 2 months prior to presentation. The mass was asymptomatic. The size of the mass was progressively increasing in the size and started to have nausea. 15 days prior to presentation she started to have abdominal pain on the right hypochonral area which was progressively increasing in intensity and was dull aching in nature with no aggravating or reliving factor. The pain was associated with nausea. No other GI symptoms could be appreciated no any constitutional symptoms. The patient came to ER for medical advice where she also underwent an Ultrasound study for the abdomen which showed, Hepatomegaly with large heterogeneous liver mass associated. Patient admitted and further evaluation by Triphasic CT liver was conducted and showed A large mass is seen involving segment 5, 4A, 4B and 8 with exophytic component. It measured 13.5 x 7 x 9.5 cm in transverse AP and craniocaudal dimensions respectively. It showed peripheral arterial enhancement central areas of necrosis and is inseparable from the abnormally thickened gallbladder wall also inseparable from a second multilobulated mass with similar pattern of enhancement seen just inferiorly measuring 9.5 x 7 x 13 cm in its transverse, AP and craniocaudal dimension respectively. This later mass appears to be invading the hepatic flexure and causing mass effect on the duodenum, pancreas with dilatation of the CBD and intrahepatic biliary duct dilatation. Small partial filling defect within segmental branches of the left portal vein which is supplying segment III. Laboratory studies showed:-

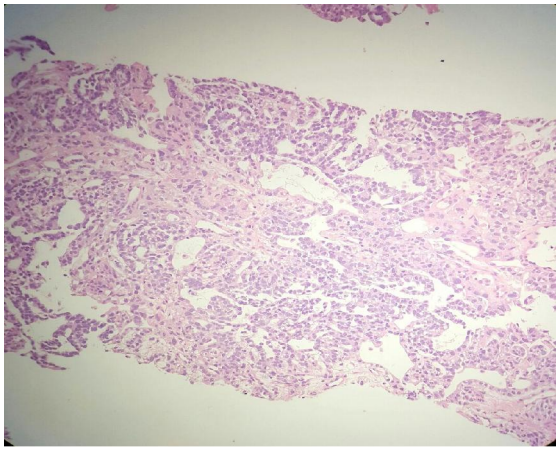
CBC: WBC 5.1, RBC 6.7, HGB 10.7, Platelate 289.

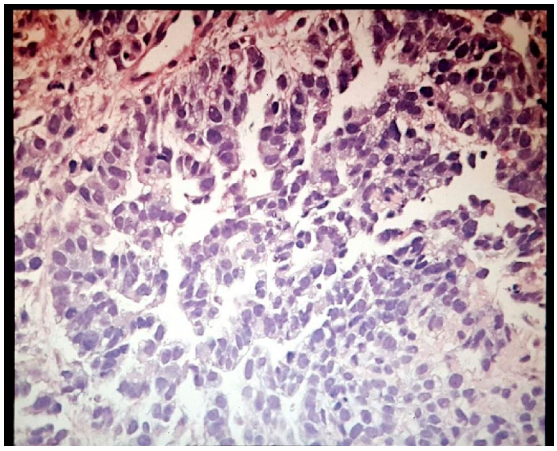
Renal Profile: Na 138, K 3.7, Cl 103, Urea 3.4, Creatinine 60.

Tumor Marker: bHCG 16, AFP 13737, LDH 350.



Thereafter, patient underwent ultrasound guided biopsy which showed a malignant neoplasm goes with yolk sac tumor and based on the Presentation, lab results and histopathology the diagnosis was a hepatoid variant of yolk sac tumor. case was explained to the patient and the treatment plan including the alternative. Patient agreed to proceed for chemotherapy with BEP protocol (bleomycin, etoposide, cisplatin). Cycle one day cycle one started on 4/07/2012 and completed day 5 cycle one on 8/07/2016 with no complication and patient discharged home with appointment to Chemotherapy day unit tow weeks after.





On 11/7/2012 patient came to the ER complaining of sever abdominal pain for this reason patient admitted and restated with fluid and pain killers then ct scan done for her which showed marked increase in the size of the ascites being massive now within the abdomen and pelvis. This is associated with scalloping of the liver surface, circumferential thickening of hypodense wall of the ascending colon and right side of the transverse with resolution of the previously described intra and extra hepatic biliary duct dilatation and mild peritoneal enhancement mainly within the upper abdomen on the right side. The overall picture is raising suspicion of billous leak peritonitis, perforation of the mass involving the gallbladder. Patient underwent exploratory laparotomy and the finding were bilious ascitis around 3.5 liter, ruptured hepatic tumor that involved right lobe. Debunking of the tumor done till we reach a grossly normal appearing liver parenchyma. Tow large drains left inside after securing hemostasis and abdomen closed. Patient shifted to ICU in stable condition. Post Op period patient developed massive PE (Saddle Shaped) and started on Heparin infusion then shifted to LMWH therapeutic dose and discharged on it. Furthermore, the chemotherapy postponed.

After resuming the cycle and completed 4 cycles follow up investigation and radiological studies showed there are two residual hypodense lesions in segment V representing residual tumor. Furthermore, There is a soft tissue well defined mass lesion at left greater omentum likely representing drop metastasis. So the decision was to proceed for TIP chemotherapy 4 cycles (paclitaxel, ifosfamide and cisplatin) on a 4 week-cycle protocol. After completion of three cycles of the TIP protocol, the investigations showed that there is improving in tumor marker results however the radiological follow up showed progression. The combined oncology meeting discussion was arising suspicion of hepatoblastoma rather than Germ cell tumor so the decision was made to proceed for 3rd line chemotherapy cisplatin, doxorubicin, etoposide, and cycophosphamide (CDEC) protocol. After the 3rd cycle of CDEC protocol, a CT scan done and showed an Interval regression of the previously seen lesions in the liver and in the peritoneum. Biomarkers including AFP 5.3, bHCG < 1, LDH 201. PET scan done and it showed that here is no metabolically active residual or recurrent tumor. Patient completed 6 cycle of CDEC. A follow up continued and CT scan done 6 months showed decrease of the size of the previously seen small hepatic lesion. MRI Abdomen done on 14/09/2014 and it showed No suspicious lesions could be identified within the liver on T2 WI, DWI or on the post contrast sequences to suggest metastasis. A six - month follow up PET scan showed No significant abnormal metabolically active could be seen to suggest active recurrent tumor or metastasis. Last radiological study was a PET CT scan and there was no FDG uptake in the chest, abdomen or pelvis. Patient seen in our clinic on 18/01/2016 and she was clinically fine and has no active medical issue.

Discussion

Yolk sac tumour (YST) follows dysgerminoma in frequency among the malignant germ cell tumours. It arises mostly in adolescents and young women through the third decade. Serum alpha-fetoprotein is elevated in most patients and the majority present with advanced stage disease.

Primary yolk sac tumour should also be considered as an alternative diagnosis to hepatoblastoma especially in children and young adult with a large liver mass and significantly raised serum alpha-fetoprotein, but in view of the nonspecific imaging features histological examination is essential for diagnosis. Although there are no specific imaging findings of primary yolk sac tumour, presence of cystic or necrotic areas and a tendency for tumour rupture are suggestive features. In the hepatoid variant

features similar to hepatocellular carcinoma occurred. However, more recently this variant has been found to occasionally produce bile in canalicular-like structures. Germ cell tumors are unique among solid neoplasms because cisplatin-based systemic therapy usually eradicates metastatic disease. This ability to cure patients with advanced disease led to evidence-based standards of care, well-defined risk-adjusted treatment algorithms, and the application of both the standards of care and the risk-adjusted algorithms in all disease stages. These achievements have resulted in an increase in the cure rate of metastatic disease from 10% in the 1960s to more than 80% today. A randomized trial comparing cisplatin, vinblastine, bleomycin, cyclophosphamide, and actinomycin-D (VAB-6) with 4 cycles of etoposide plus cisplatin (EP×4) revealed equivalent complete remission rates and EP×4 had less toxicity.Other RCT examined standard regimen of 4 cycles of BEP (bleomycin, etoposide, cisplatin; BEP×4) in comparison with BEP×3 and showed that the two regimens had equivalent cure rates and minimal pulmonary toxicity. Up to 30% of patients with advanced germ cell tumors will either relapse or fail to achieve a complete remission and will require salvage chemotherapy. A thorough review of this clinical scenarioshas recently been published. At present, one of the major salvage approachis conventional-dose chemotherapy (CDCT). CDCT regimens incorporate cisplatin, ifosfamide, and either vinblastine (VeIP) or paclitaxel (TIP). In two series of patients who received initial salvage chemotherapy, the durable complete remission rate for VeIP was 23% and for TIP it was 63%. As a result, TIP has become the preferred CDCT regimen.

Surgery should be considered in all patients with advanced nonseminomatous germ cell tumors whose markers normalize. Except under certain circumstances with persistent elevation or an increase in the AFP and/or hCG level implies residual viable disease and requires treatment with second-line chemotherapy. The National Comprehensive Cancer Network guidelines and all germ cell tumor experts recommend retroperitoneal lymph node dissection (RPLND) after chemotherapy in patients with advanced nonseminomatous germ cell tumors for any residual retroperitoneal nodes > 1 cm.

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