Cystic Presentation of Primary Hepatic Angiosarcoma

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Case Report

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Abstract

Primary hepatic angiosarcoma is a rare malignancy arising from the endothelial cells. They account for 1% of all primary liver tumors. They are aggressive tumors and patients usually present with distant metastasis. Surgical resection is the mainstay of treatment, however recurrences are common. The overall survival of patient with hepatic angiosarcoma is only five months without any treatment. Addition of adjuvant chemotherapy with surgery seems to prolong the overall and disease-free survival. Their diagnostic dilemma is due to the fact of non-specific imaging findings mimicking other hypervascular liver tumors. Angiosarcomas are solid tumors arising from the endothelial cells. Cystic type of angiosarcomas are rare and those arising from the liver have not been reported. We are presenting a case report on a cystic primary hepatic angiosarcoma.

Key Message

With an incidence of 1% and non-specific clinical and imaging findings, a strong clinical suspicion is required to diagnose a case of primary hepatic angiosarcoma.

Introduction

Primary hepatic angiosarcoma (PHA) is an uncommon malignant tumor with an incidence of 1% of all hepatic tumors. There is no definitive etiology for PHA, however its association with exposure to polyvinyl chloride, arsenic and thorotrast. Patients present with non-specific symptoms and there are no definitive imaging findings to clinch the diagnosis. The median survival of patients diagnosed with PHA is on 5 months without any treatment. Surgery with an intention to achieve R0 resection is the mainstay of the treatment. The low overall survival can be attributed to the aggressive disease and late presentation of patient with distant metastasis. We are reporting a case of PHA with a cystic presentation which has not been reported in literature.

Case History

Our patient is a 65-year-old female who presented to our outpatient clinic with complaints of diffuse abdominal pain of one month duration. Neither was there a history of abdominal distension, nausea and vomiting nor any history of loss of appetite or weight. She was evaluated with a contrast enhanced CT scan of the abdomen which revealed a 13.3 x 10.7 x 11.3 cms multiloculated cystic lesion in segment VIII of liver extending into segment IVA and VII and up to the sub-capsular aspect of liver with multiple enhancing septations and few areas of mural nodularity (Fig 1). The said lesion is seen causing a mass effect on adjacent hepatic venous channels.

The biochemical parameters of the patient pre-operatively are mentioned in Table 1.
<table>
<thead>
<tr>
<th>PARAMETER</th>
<th>VALUE</th>
<th>REFERENCE VALUE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin</td>
<td>12.0</td>
<td>12 - 15 gm/dl</td>
</tr>
<tr>
<td>Total Count</td>
<td>9629</td>
<td>4000 - 11000 cu mm</td>
</tr>
<tr>
<td>Platelet</td>
<td>260000</td>
<td>150000 - 400000 cu mm</td>
</tr>
<tr>
<td>INR</td>
<td>1.18</td>
<td></td>
</tr>
<tr>
<td>ESR</td>
<td>23</td>
<td>0 - 22 mm/hr</td>
</tr>
<tr>
<td>RBS</td>
<td>304</td>
<td>70 - 160 mg/dl</td>
</tr>
<tr>
<td>Urea</td>
<td>18.4</td>
<td>10 - 50 mg/dl</td>
</tr>
<tr>
<td>Creatinine</td>
<td>0.5</td>
<td>0.6 - 1.1 mg/dl</td>
</tr>
<tr>
<td>Total Bilirubin</td>
<td>0.8</td>
<td>0.3 - 1 mg/dl</td>
</tr>
<tr>
<td>Direct Bilirubin</td>
<td>0.2</td>
<td>0.1 - 0.47 mg/dl</td>
</tr>
<tr>
<td>Indirect Bilirubin</td>
<td>0.6</td>
<td>0 - 0.3 mg/dl</td>
</tr>
<tr>
<td>SGOT</td>
<td>20</td>
<td>10 - 40 U/L</td>
</tr>
<tr>
<td>SGPT</td>
<td>23</td>
<td>10 - 37 U/L</td>
</tr>
<tr>
<td>ALP</td>
<td>90</td>
<td>39 - 117 U/L</td>
</tr>
<tr>
<td>Total Protein</td>
<td>6.9</td>
<td>6 - 8 gm/dl</td>
</tr>
<tr>
<td>Albumin</td>
<td>4.0</td>
<td>3.5 - 5 gm/dl</td>
</tr>
<tr>
<td>HBS Ag</td>
<td>Non-reactive</td>
<td></td>
</tr>
<tr>
<td>HCV</td>
<td>Non-reactive</td>
<td></td>
</tr>
<tr>
<td>Hydatid IgG</td>
<td>Positive - 17.4</td>
<td>&gt;11</td>
</tr>
</tbody>
</table>

**Tumour Markers**

<table>
<thead>
<tr>
<th>Marker</th>
<th>Value</th>
<th>Reference Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>AFP</td>
<td>6</td>
<td>&lt;7.22 IU/ml</td>
</tr>
<tr>
<td>CA 19.9</td>
<td>&lt;2.00</td>
<td>0 - 30.90 U/ml</td>
</tr>
<tr>
<td>CEA</td>
<td>2.44</td>
<td>&lt;3.00 ng/ml</td>
</tr>
</tbody>
</table>

On physical examination her performance status was found to be fair (ECOG 0). Her systemic examination was unremarkable. At this time the differential diagnoses were Biliary cystadenoma with
internal haemorrhage, hemorrhagic hepatic cyst, hydatid with internal haemorrhage. She was empirically started on oral albendazole. After obtaining Pre-anesthetic clearance and optimizing the patient, she was taken up for en-bloc excision of liver cyst.

On table, a 15 x 10 cms cystic lesion was seen replacing segments IVa, VII and VIII with adhesions to the right hemidiaphragm (Fig 2). Feeding vessels from right anterior sectoral and segment 4 vascular pedicels were identified and controlled. Both the right and middle hepatic veins were splayed by the tumour. The procedure ended up being a modified hepatectomy of segments IVa, VII and VIII along with a part of the right hemidiaphragm. Her intra-operative and post-operative periods were uneventful and she was discharged in a hemodynamically stable on the sixth post-operative day.

The histopathological examination revealed a multiloculated lesion filled with brownish material and focal solid area and large areas of hemorrhage. Microscopy showed sheets of spindle shaped pleomorphic cells with thin-walled vascular channels interspersed. This was suggestive of a malignant spindle cell neoplasm possible angiosarcoma with the closest resection margin being 0.4mm away. It was decided to start her on adjuvant chemotherapy with Gemcitabine (1200) and Docetaxel (100mg). She has remained asymptomatic and disease free for a period of three years since the procedure. The patient has given an informed consent for publishing this data.

Discussion

Primary hepatic angiosarcoma (PHA) is a rare malignant tumour arising from the endothelial cells in the liver which accounts for 1% of all primary hepatic tumours with males being more affected than females (4:1) \(^1,2,3\). Most common primary hepatic malignancies intrahepatic cholangiocarcinoma followed by hepatocellular carcinoma, with PHA being third in line \(^1,2\). Median survival time of patients diagnosed with PHA is only 5 months \(^3\).

The difficulty in diagnosing hepatic angiosarcoma is due the lack of any specific tumour markers and specific imaging findings. 75% of the tumours have no definitive etiology \(^4\). It is associated with malignant transformation usually seen in relation to exposure to polyvinyl chloride, arsenic, Thorotrast and radiation \(^1,3,4\).

Patients usually present with non-specific symptoms such as weight loss, abdominal pain, fatigue, etc \(^5\). Some present with rupture and intraperitoneal haemorrhage where the first choice to stop the bleeding is Transarterial embolization \(^6\).
Most hepatic angiosarcomas are hypoattenuating with some of them showing ring enhancement, necrosis and hemorrhage. There are no definitive findings on imaging to diagnose PHA. Its pleomorphic histology produces various patterns of tumor enhancement. Frequently, contrast enhanced imaging shows hypoattenuation reflecting raised tumor vascularity, although there have been cases reported with focal lesions on delayed imaging. According to Byrnes et al, interval growth and rim enhancement of arterially enhanced nodules of size <2cm are associated with malignancy.

The definitive diagnosis of PHA is on histopathological examination. Gross pathology usually reveals two patterns of growth; a large solitary mass, multi nodular or multifocal lesions, mixed patterns of dominion mass with smaller nodules or an infiltrating micro nodular tumor. Immunohistochemical markers like CD31, CD34 and factor VIII are positive in PHA.

The mortality associated with PHA is usually high due the late presentation of the patients in an unresectable stage.

Surgical resection of the liver remains the mainstay of treatment, since there is no effective chemotherapy regimen suggested in the literature, although they are being given as palliation. In literature, surgical resection with adjutant chemotherapy has increased the median survival to 40.5 months.

Literature suggests treatment of PHA has poor outcomes. Good prognostic factors seem to be single tumor, small tumor size, lack of metastases, low grade lesion and negative surgical margins. Metastasis of PHA is usually seen to lungs, spleen and bone and are seen early in the disease course.

Conclusion

PHA presents with vague symptoms which progresses very rapidly, to which the patient succumbs usually. Sensitivity of imaging is unreliable and hence a high clinical suspicion is required to diagnose this pathology.

Declarations

Competing Interests

The authors declare that there are no competing interest of personal or financial nature.

Author's Contributions

J and A – prepared the manuscript. J,A,G – reviewed the literature. J- prepared the images. All authors reviewed and checked the article.

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References


5. Yang KF. Primary Hepatic Angiosarcoma: Difficulty in Clinical, Radiological, and Pathological Diagnosis.


Figures
Figure 1

CT image showing a heterogenous lesion in the right lobe of liver
Figure 2

Cystic lesion seen arising from the right lobe of liver