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An atypical presentation of Liposarcoma: Primary involvement of the liver with secondary metastatic seeding

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An atypical presentation of Liposarcoma: Primary involvement of the liver with secondary metastatic seeding

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Abstract

Metastatic liposarcoma to the liver is rare, with primary liposarcomas of the liver even more so. There have only been about a dozen cases of primary liver liposarcoma reported in the literature. The knowledge of the clinical course, management, and prognosis are limited. Here, we present a well-differentiated primary liposarcoma of the liver originating in the porta hepatis of a 61-year old male patient that extended into the retroperitoneum, causing mass effect on the inferior vena cava (IVC).

Introduction

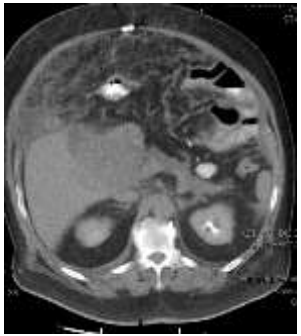
- Soft tissue sarcomas are a rare malignancy compromising about 1% of all found in adults
- Liposarcoma are a subclass of soft tissue sarcomas arising from precursor adipocytes
- Normally Liposarcoma are found in the retroperitoneum and extremities
- Involvement of the liver is very rare and usually from metastases rather than a primary focus.
- Only a dozen cases of primary liver Liposarcoma have been reported in literature review, with no gender association and age distributions ranging from 26 months to 86 years
- Little is known about management, clinical course and prognosis of such cases

Case Description

- Pt is 61-year old male presenting to the ED from outside hospital with a10-day history of SOB and abdominal distention. Initial imaging and labs were done at OSH (Figure 1)
- **PMH**: gastric bypass and previous EtOH abuse
- **ROS**: *endorsed* difficulty with taking deep breaths, urinating, and bowel movements, *denied* unexplained weight loss, night sweats, or history of ascites
- **PE**: abdominal distension and tenderness

Work Up

- Hep screen, AFP, CEA, and CA19-9 were *negative*
- AST, ALT, total bili, and alkaline phos were WNL
- **CT**: pleural effusion with atelectasis, large amounts of ascites with mesenteric stranding, and a 7cm mass of unknown etiology adjacent/medial to the liver



- **Figure 1** shows CT chest/abdomen/pelvis w/ contrast from outside hospital demonstrating mass and ascites adjacent and medial to liver

Outcome and Follow-Up

- Surgical intervention not recommended
- Plan for outpatient chemotherapy with ifophosphamide and doxorubicin
- At first appointment, pt was transferred to the hospital for management and chemo initiation
- Workup included CTPE, ECHO, and thoracentesis which demonstrated bilateral PE, new onset HFrEF, and parapneumonic effusion
- Considered a poor candidate for palliative chemotherapy, with an overall poor prognosis
- Goals of care discussed with hospice care recommended
- Therapeutic thoracentesis done and pleurX catheter placed prior to d/c
- **EGD** *negative* for portal HTN
- **Paracentesis** and **thoracentesis** *negative for malignant cells*
- **CT-guided biopsy** consistent with well-differentiated liposarcoma
- **IR-guided biopsy** demonstrated de-differentiated liposarcoma, FNCLCC grade 2
- **Colonoscopy** found three polyps demonstrating tubular adenoma
- Diagnosis: metastatic primary liposarcoma of the liver

Discussion

- Primary Liposarcoma of the liver is very rare →little data out to support management strategies
- It is important to continue to gather data about outcomes of patients, as management strategies should be implemented only if they can improve outcomes.
- In this case, patient was not a candidate for surgical resection / radiation due to poor tolerance by the liver and extensive and advanced disease
- Ultimately started on OP chemotherapy, but had rapid deterioration needing a goals of care discussion
- Currently suggested management includes: curative and aggressive surgery with negative margins, regular sonographic examinations in as little as 12-week intervals
- Without treatment prognosis is very poor
- If the outcomes do not look favorable hospice and palliative care may be implemented earlier

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