Metastatic Leiomyosarcoma Presenting as Polycystic Liver Disease

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This is a case of a 39-year-old male developing intra-abdominal leiomyosarcoma (LMS) with hemorrhagic liver metastasis masquerading as Polycystic Liver Disease (ADPLD). He had been mistakenly diagnosed with ADPLD twenty-one months prior to biopsy. This is an atypical presentation in an already rare disease. Patient’s with ADPLD containing complex or hemorrhagic cysts should be considered for dynamic MRI to rule out metastatic disease mimicking ADPLD.

A 39 year-old-male who presented with progressive stabbing and constant abdominal pain. It was associated with jaundice, poor appetite, night sweats and 40 lbs weight loss. A non-contrast CT abdomen/pelvis showed hemorrhagic liver cyst with possible abscess.

He was diagnosed with polycystic liver disease twenty-one months earlier by CT scan. Some nodularity around cysts was noted and dynamic MRI was ordered, but the patient was lost to follow up.

Dynamic MRI demonstrated evidence of a pancreatic arterial enhancing mass concerning for neuroendocrine tumor. Multiple scattered nodular arterial enhancing lesions with numerous complex hemorrhagic, loculated hepatic cysts were seen consistent with metastatic disease.

Retroperitoneal lymph node was biopsied and demonstrated spindle cell malignancy compatible with leiomyosarcoma.

No palliative chemotherapy was pursued. He was discharged on hospice.

Leiomyosarcoma is a malignant smooth muscle tumor that can originate from virtually any part of the body. The most common sites affected are the retroperitoneum and the peritoneal cavity. The peak incidence of leiomyosarcoma is in the sixth and seventh decades.

According to the World Health Organization, LMS of the gastrointestinal tract is so rare that there is no significant data on demographic, clinical, or gross features of the tumor. One study found that over twelve-year period 56 cases of leiomyosarcoma were identified in the GI tract. Liver metastases occurred in 27% of the time.

The atypical presentation of LMS with advanced necrotic liver metastasis can be explained by its propensity to cause necrotic, liquefaction, and cystic change in the liver.

LMS is a fascicular spindle cell neoplasm, and tumor cells have brightly eosinophilic cytoplasm and cigar-shaped nuclei. LMS generally lack KIT and CD34 expression but reliably demonstrate high levels of smooth muscle actin and desmin.

Despite frequent tumor recurrence, the long-term outcome after liver resection for hepatic metastases from LMS is superior to that after chemotherapy and chemoembolization.

We recommend dynamic MRI of the liver with any patient that has suspicious radiographic features when evaluating patients with ADPLD.