Primary Malignant Mixed Mullerian Tumor: An Uncommon Liver Cyst

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Introduction

- Carcinosarcoma, or malignant mixed Mullerian tumor (MMMT), is a biphasic malignant neoplasm consisting of epithelial and non-epithelial components. These tumors typically rise from the female genital tract.
- Primary extragenital MMMTs are exceedingly rare.

Case

- 71 yrs old woman presented with 3 mos history of diffuse abdominal pain and distension.
- Physical examination showed a distended abdomen with liver edge palpable 4 cm below the coastal margin.
- Laboratory data: WBC 14 k/uL, ALT 50 U/L, AST 86 U/L, AP 388 U/L, and total bilirubin 0.3 mg/dL, CA 125 481 U/mL, AFP 42 ng/mL, CA 19-9 14 U/mL, and CEA < 1.2 ng/mL.
- CT abdomen and pelvis (Figure 1A and 1B)
- She underwent drain placement into the liver cyst .Fluid cultures and cytology showed no infection or malignancy.
- Laparoscopic abdominal exploration demonstrated omental caking and multiple peritoneal nodules.

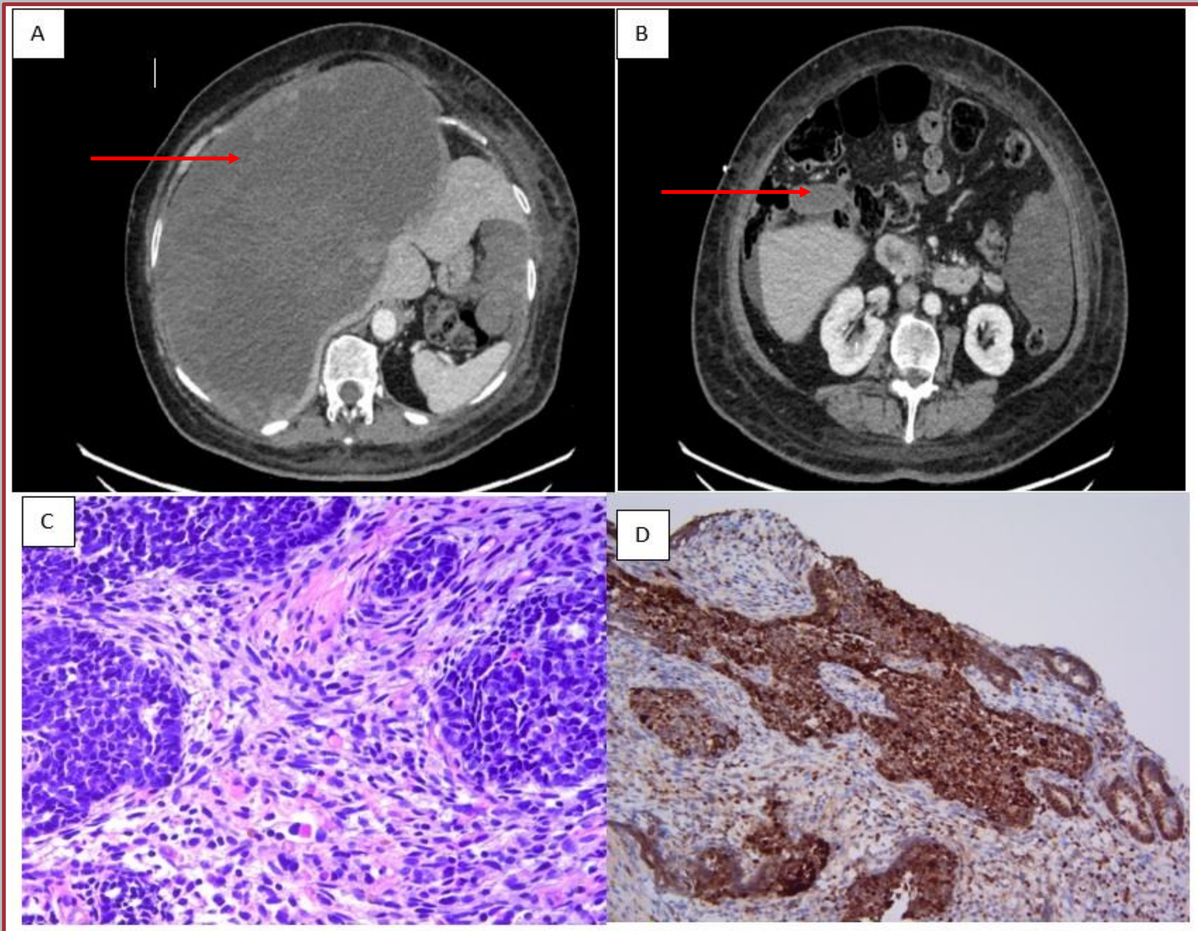

Figure 1A and B) CT abdomen and pelvis with multiloculated cystic mass in the liver measuring 22 cm with septation and cystic lesion in the left peritoneum adjacent to the ascending colon measuring 15 cm 1C)Islands of malignant epithelial cells are separated by a malignant cellular stroma with rhabdomyosarcomatous differentiation (H&E, 100x) 1D) Pax8 immunostain is positive in the epithelial components (100x)..

- Omental nodule biopsies (Figure 1C and 1D)
- CT chest, colonoscopy, and transvaginal ultrasound were unremarkable.
- Diagnosis of primary peritoneal carcinosarcoma was made.
- Carboplatin and Paclitaxel were started. She was not deemed a candidate for cytoreductive surgery and opted for hospice.

Discussion

- Primary peritoneal carcinosarcomas mostly occur in the pelvic peritoneum, followed by serosal surface of the colon, retroperitoneum, and omentum
- Extragenital carcinosarcomas are rare.
- Carcinosarcomas of extragenital sites are thought to arise from derivatives of the coelomic epithelium.: Müllerian duct remnants, secondary Müllerian system, or pre-existing foci of endometriosis.
- Due to a common embryonic origin of the ovary and peritoneum, they have much histologic similarity. Most cases occur in women above the age of 40
- Complete cytoreduction surgery and systemic chemotherapy is the mainstay of treatment.
- These tumors have poor prognosis with average survival rate between 11-17 months.