Carcinosarcoma
WHAT SHOULD I KNOW?

BACKGROUND
The uterus and the ovaries have two predominant tissue components – the epithelium and the stroma. In the ovary, the epithelium can be thought of as the cover or shell and the stroma is the meat within. The epithelium of the uterus is predominantly a thin lining in the middle surrounded by the thick stroma and smooth muscle. In both the ovary and the uterus, the most common cancers arise from the epithelial component. In rare cases, a tumor will display a mixture of malignant components from both the epithelium and the stroma. These rare tumors are called malignant mixed müllerian tumors (MMTs), or more commonly today a carcinosarcoma. Carcinosarcomas represent less than 5% of ovarian cancers and less than 10% of uterine cancers, and staging differs based on the site of origin. Regardless of site of origin and stage, carcinosarcomas are highly aggressive tumors that have often metastasized by the time of diagnosis. Frequent metastatic sites are the lymph nodes, adjacent pelvic and peritoneal structures, and lungs. Due to the rarity of these tumors, it is difficult to study them or develop novel therapies and treatment strategies. There does not appear to be any established hereditary link to most carcinosarcomas.

SCREENING
Currently there is no standardized screening for carcinosarcoma. You should seek medical evaluation from a gynecologist if you are experiencing abnormal uterine bleeding, pelvic pain or pressure, bloating, constipation, and early satiety.

TREATMENT
Surgery: Hysterectomy, removal of fallopian tubes and ovaries, lymph node sampling, and surgical excision of any metastatic carcinosarcoma is a key component of therapy.
Chemotherapy: Chemotherapy is typically recommended for all stages regardless of site of origin due to high risk of recurrence. Regimens often include multiple drugs. Common drugs are carboplatin, paclitaxel, ifosfamide.
Radiation: In many cases pelvic radiation is prescribed after initial diagnosis and surgery in conjunction with chemotherapy. Radiation may be used palliatively as well.
Targeted Therapy: Larotrectinib or entrectinib for NTRK gene fusion containing tumors.
Immunotherapy: Pembrolizumab or nivolumab may be offered in tumors classified with high tumor mutational burden or other genetic markers suggesting potential benefit from immune therapy.
Hormonal Therapy: Some carcinosarcomas may overexpress estrogen or progesterone receptors and hormonal therapy could be an adjunct in these cases.

QUESTIONS YOU SHOULD ASK
Am I eligible for any clinical trials? What stage is my cancer? Is my doctor a gynecologic oncologist? Should I get a second opinion? Can genetic testing be done on my tumor?