Mucinous Ovarian Cancer

WHAT SHOULD I KNOW?

DESCRIPTION OF THE TUMOR/CANCER TYPE

Mucinous ovarian cancers are rare, accounting for 3-5% of all primary ovarian cancers. The median age of diagnosis is 55, but can also be seen in younger women. The tumor tends to be involving one ovary (unilateral), which allows for the option of fertility sparing approaches to surgery in patients who desire fertility. Because mucinous tumors can more commonly arise from the gastrointestinal tract (i.e., colon, stomach, appendix), it is important to make sure the mucinous tumor originated in the ovary and is not a spread (metastasis) from another site such as the appendix. Other tests, such as using a scope to look in your stomach (EGD) and colon (colonoscopy), need to be done to rule out a different source of the cancer. Under the microscope, pathologists can use features such as the association of the cancer cells with other benign or low malignant tumor mucinous cells which would be more indicative of an ovarian primary tumor. Special stains can be performed on the cancer cells to look for markers that help differentiate between primary mucinous ovarian cancers versus metastasis from other sites. The pattern of the primary ovarian cancer under the microscope can also be associated with aggressiveness of the cancer and risk of it coming back. Confluent or expansive appearance of the cancer has lower risk of recurrence, whereas infiltrative-destructive appearance is associated with a higher risk of recurrence.

SIGNS & SYMPTOMS

Mucinous ovarian cancers are often large tumors that can cause a change in how clothes fit and sometimes a mass or lump is palpable. Mucinous tumors can also cause pelvic pain, filling up easily with eating, or nausea.

SCREENING

There is currently no effective screening test for ovarian cancer.

TREATMENT & FOLLOW-UP

This section is intended to clarify basics and showcase how treatment and follow-up may be different for these rare types of cancer.

Role of Surgery

Mucinous ovarian cancers are often large tumors that can impact the surgical approach used to remove them. It is important to remove them intact. Because mucinous ovarian cancers can impact young women and they are typically limited to one ovary, fertility sparing surgery and staging can be done to preserve fertility for patients who are not through with childbearing. This approach to surgery typically entails some fluid being placed in the pelvis to look for cancer cells (pelvic washings), removal of the mass that can be part or all of the ovary depending on how much of the ovary is involved, removal of lymph nodes in the pelvis and up higher near the aorta, removal of the omentum (a piece of fat that hangs like an apron off of the bowel), and biopsies of the lining for the abdomen and pelvis. The appendix, if still in place, is also typically removed in mucinous cancers to be sure the mass is not originating from the appendix.

Role of Chemotherapy

Fortunately, the majority of mucinous ovarian cancers are stage I and do not require any therapy after surgery. However, mucinous cancer that have spread to other areas (stage II-IV) or cancer that recurs typically require therapy after surgery. Mucinous ovarian cancers are not very responsive to chemotherapy. Recommendations for treatment include therapies used for other types of ovarian cancer (paclitaxel/carboplatin/bevacizumab) versus therapies used in gastrointestinal cancers (5-fluorouracil/leucovorin/oxaliplatin plus/minus bevacizumab or capecitabine/oxaliplatin plus/minus bevacizumab). Research studies performed comparing the regimens did not show benefit of one over the other because of small numbers of patients given the rare tumor type.

Targeted Therapy Options

Molecular testing done on mucinous ovarian cancer detected several findings. A significant number of mucinous cancers have a KRAS/NRAS or p53 mutation. Twenty-six percent of these cancers have amplification of ERBB3. Other mutations, such as RNF43, ARID1A, and PIK3CA/PTEN, were also identified. All of these have potential targeted therapies that could be available in a research trial especially for patients with advanced disease not responsive to therapy or recurrent cancer. The rarity of homologous recombination deficiency (tumor DNA repair deficiency) in mucinous ovarian cancer makes it less likely that they will be responsive to PARP-inhibitor therapy.
**Immunotherapy Options**
Mismatch repair deficiency (difficulties in DNR repair in tumor) is a marker evaluated for possible response of cancer to immunotherapy and is rarely found in mucinous ovarian cancer. Therefore, the benefit of immunotherapy in mucinous ovarian cancer is unknown.

**Hormonal Therapy**
Estrogen receptors were expressed in some mucinous ovarian cancers, so hormonal therapy may be an option for some patients.

**QUESTIONS YOU SHOULD ASK YOUR CARE TEAM ABOUT YOUR TREATMENT PLAN & FOLLOW-UP CARE**
If you are not already being treated by a gynecologic oncologist, consider seeking a second opinion.

- What is the best approach for my surgery?
- Is there an option to preserve my ability to have children with this cancer?
- What is the stage of my cancer?
- Do I need any treatment after surgery? If so, is a research trial an option?