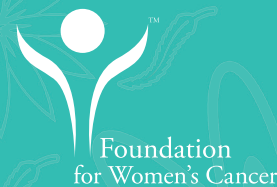




Clear Cell Carcinoma of the Ovary

WHAT SHOULD I KNOW?



DESCRIPTION OF THE TUMOR/CANCER TYPE

Clear cell carcinoma of the ovary is the second to third most common type of ovarian cancer accounting for 5-11% of all ovarian cancers. It is equally as common as endometrioid type, and both histologic types are associated with endometriosis. Endometriosis is a risk factor as well as a precursor lesion for clear cell carcinoma. It is seen in 50% of these cancers and can be unexpectedly found at surgeries done for endometriosis. Clear cell carcinomas are seen in younger women, tend to be involving one ovary, and the majority are early-stage disease. Clear cell carcinomas can be associated with a lot of scarring and adhesions at the time of surgery. CA125 (a blood test marker for ovarian cancer) is typically not as elevated in clear cell carcinoma as other types of ovarian cancer. Clear cell carcinomas have been associated with elevated calcium levels and increased risk of developing blood clots.

SIGNS & SYMPTOMS

Just like other types of ovarian cancer, clear cell carcinomas can be associated with pelvic pain, abdominal bloating, and change in bowel function. Because of its association with endometriosis and impact in younger women, symptoms such as abnormal vaginal bleeding, pain with sex, painful periods, and changes in urinating can also be seen.

SCREENING

As with other types of ovarian cancer, there is currently not a good screening test for clear cell carcinomas of the ovary. However, some clear cell carcinomas of the ovary are associated with Lynch syndrome (a hereditary cancer syndrome) which can be detected by genetic testing, so genetic testing for patients with ovarian cancer is recommended.

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

Having pathology look at the mass under the microscope at the time of surgery (frozen section) to make the diagnosis of cancer

may allow surgical staging to happen at the time of removal of the pelvic mass. Staging typically involves removal of the ovary and fallopian tube, placing fluid in the pelvis to look for cancer cells, removal of lymph nodes down in the pelvis and up higher near the aorta, biopsies of the lining of the abdomen and pelvis, and removal of the omentum (a piece of fat that lays on the bowel). If the clear cell carcinoma patient does not want more children, removal of the uterus, both fallopian tubes and ovaries, as well as the other staging biopsies, are performed. Fortunately, clear cell carcinoma is often diagnosed at an early stage, but a staging procedure is important to confirm the stage which helps determine the amount of therapy after surgery that is needed.

Role of Chemotherapy

Clear cell carcinoma of the ovary is less responsive to chemotherapy than other types of ovarian cancer. Because it can behave in a more aggressive manner, therapy after surgery with chemotherapy is usually recommended. Typically, paclitaxel and carboplatin chemotherapy is recommended for stage I disease. For patients with stage II-IV disease, treatment with paclitaxel and carboplatin chemotherapy for six treatments versus paclitaxel/carboplatin/bevacizumab (a targeted therapy that impacts blood vessel formation in cancer) for six treatments followed by maintenance therapy with bevacizumab are preferred options.

If clear cell carcinoma of the ovary recurs, it can be difficult to treat since it is less responsive to chemotherapy. There are several options available depending on the timing of the recurrence. If cancer returns over six months after completing treatment, it may be more responsive to chemotherapy and there are several options of combination chemotherapies available. If the cancer returns quickly, it may be resistant to chemotherapy and options for other types of therapies such as radiation, targeted therapies, immunotherapies, or participation in a clinical trial may be better options for treatment.

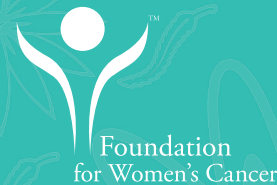
Radiation Therapy

There is some data to suggest that clear cell carcinoma of the ovary may be more responsive to radiation therapy. If a clear cell carcinoma returns in a single area, focal radiation may be used for treatment.



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Targeted Therapy Options

As mentioned above, bevacizumab (a drug that targets how blood vessels in cancer are different than other blood vessels) can be used for treatment in advanced stage or recurrent clear cell carcinoma of the ovary. Molecular testing of clear cell carcinoma may identify potential targets for therapy. Targeted therapy options may be available for patients with recurrent disease through a clinical trial.

Immunotherapy Options

Six-10% of clear cell carcinomas of the ovary have difficulty repairing DNA which is detected by testing for microsatellite instability (MSI). Tumors that are MSI-high may be responsive to immunotherapy. Tumor testing of clear cell carcinoma can be done. If the tumor expresses PD-L1 or has a high tumor mutational burden, it may respond to treatment with immunotherapy.

Hormonal Therapy

The majority of clear cell carcinomas of the ovary are negative for estrogen receptors so the role of hormonal therapy is uncertain.

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 stage of my cancer?

Do I require any treatment after surgery?

Am I being referred to genetic counselling because of my diagnosis?

Are other tests being done to my tumor to see if I would benefit from other types of therapy if treatment after surgery is needed?



Society of Gynecologic Oncology

The Foundation for Women's Cancer (FWC) is a 501(c)3 nonprofit organization dedicated to supporting research, education, and public awareness of gynecologic cancers. The FWC is the official foundation of the Society of Gynecologic Oncology. Learn more at foundationforwomenscancer.org.