Sex Cord Stromal Tumors

WHAT SHOULD I KNOW?

Foundation for Women's Cancer

DESCRIPTION OF THE TUMOR/CANCER TYPE

Ovarian sex cord stromal tumors (SCSTs) comprise roughly 7% of all >21,000 ovarian cancers diagnosed annually in the United States. These tumors arise from the cells that surround and support germ cells (i.e., eggs). They tend to affect women younger than 40 years. Many secrete proteins that can be measured in the bloodstream known as 'tumor markers,' such as inhibin B, estrogen (E2), testosterone (T), alpha fetoprotein (AFP). Benign sex cord stromal tumors include fibromas and thecomas. These benign tumors can sometimes mimic cancerous tumors by producing ascites (abnormal fluid in the abdomen) as part of Meigs' syndrome or even bowel obstruction (sclerosing peritonitis). There are several subtypes of malignant SCST:

|--SUBTYPE--| |----------TUMOR MARKER----------|

	Inhibin B	E_2	T	AFP	Mutation
Granulosa cell, adult (95%) and juvenile (5%)	+	+ -	+ -	-	*FOXL2*
Sertoli -Leydig	+ -	+ -	+ -	+ -	*FOXL2, DICER1*
SCTAT*	-	+	-	-	*STK11*
Steroid cell tumors	+ -	+ -	+ -	NR***	
Sex-cord stromal tumor, NOS**	+ -	+ -	+ -	+ -	

*sex cord tumor with annular tubules
**not otherwise specified
*** not reported

Fibrosarcoma is an extremely rare and aggressive subtype of SCST.

SIGNS & SYMPTOMS

SCSTs are frequently hormonally active. Tumors which secrete estrogen may produce abnormal vaginal bleeding after menopause, amenorrhea (the absence of vaginal bleeding) in pre-menopausal patients, a secondary endometrial cancer or pre-cancer, or precocious (early) puberty in children. Those which secrete testosterone may cause virilization, including male pattern hair growth, acne, and deepening of the voice. Granulosa cell tumors are the most common subtype of SCST. Larger tumors may be associated with abdominal pain or swelling.

SCREENING

There are no good screening tests for this condition. Patients with Peutz-Jeghers syndrome have an increased risk of Sertoli tumors and SCTATs, so this should be considered upon diagnosis of a pelvic mass in patients with this condition.

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

Fertility-sparing surgery can often be performed safely if it appears disease is confined to one ovary. This could include the removal of one ovary and fallopian tube, with biopsies of the omentum and lining of the abdomen, and sometimes the lining of the uterus. Because it is rare for this disease to spread to lymph nodes, lymph node removal can usually be omitted unless the lymph nodes appear obviously enlarged. Either an open (using one large incision) or laparoscopic (using several small incisions) approach may be suitable. If fertility-sparing is offered, completion staging can be performed after childbearing is complete. Surgery establishes the diagnosis, allows the tumor to be staged (extent of spread at the time of diagnosis), and is also the first step in treatment.

Role of Chemotherapy

Patients with low-risk disease (confined to the ovaries, well-differentiated) can be treated with surgery alone. If the tumor has ruptured or has certain aggressive features under the microscope (moderately to poorly differentiated, heterologous elements), or presents at stage II-IV, chemotherapy is often recommended consisting of 4-6 cycles of

carboplatin + paclitaxel

OR

etoposide + cisplatin

OR

bleomycin + etoposide + cisplatin

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In some cases, hormone therapy can be used to treat SCSTs.

Follow-Up

Upon remission, patients with SCSTs should undergo reassessment (clinical exam and biomarkers) every 6-12 months if low-risk and every 4-6 months if high-risk. Some SCSTs (e.g., granulosa cell tumors) have been known to recur many decades after initial diagnosis; therefore extended or even lifelong surveillance may be beneficial. Stage (extent of spread at the time of diagnosis) is the most important factor in outcome. Among the most common histologies, five-year survival is >90% for stage I disease, but <50% for stage III/IV disease.

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.

Does my cancer have a tumor marker?

What was my stage at diagnosis?

Should I be evaluated by fertility specialist?

Are there any clinical trials available for my disease?

SOURCES & MORE INFORMATION

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Siegel RL, Miller KD, Fuchs HE, Jemal A. Cancer Statistics, 2021. CA Cancer J Clin. 2021 Jan;71(1):7–33.

SGO

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.