



Patient Report

Sex Cord Tumors

A Report by
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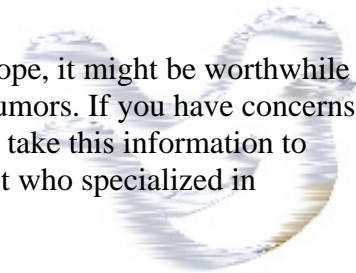
To introduce the topic, I suggest the article *Peutz-Jeghers Syndrome* by Thomas McGarrity, M.D. et al. published in **American Journal of Gastroenterology** Vol. 95, No. 3, 2000. The authors give an up-to-date description of the clinical effects of PJS. There is nearly a page devoted to genital tract tumors associated with PJS. They describe different types of tumors and their presenting symptoms. A quote: "This lesion (sex cord tumors with annular tubules) has been found in almost all female patients with PJS in whom the ovaries have been examined."

I also suggest reading the article *Very high risk of cancer in familial Peutz-Jeghers syndrome* by Giardiello et al. published in **Gastroenterology** Vol. 119, No. 6. In table 4 the projected cumulative risk rate of cancer in patients with PJS from ages 15 to 64 is forecast. The cumulative risk for ovarian cancer is 21%, cervical cancer is 10% and testicular cancer is 9%. In an e-mail I wrote to the listserv on January 31, 2002, I questioned the accuracy of these predictions because they are based on a study population of only 210 patients. Table 5 includes information from many PJS case reports. Included are the following data: Ovarian tumors were reported in 53 females (49 had sex cord tumors), cervical cancer in 28 women (22 cases of Adenoma malignum) and testicular tumors reported in 9 males (all Sertoli cell tumors).

We don't really know the number of people with PJS, but it was estimated to be 600 in 1980. The syndrome was described by Dr. Jeghers (along with Dr. McKusick and Dr. Katz) in 1949. Estimates of incidence rates range from 1 in 8,000 to 1 in 280,000. It is difficult to determine just what the numbers mean for those of us with PJS. If we don't know how many people have PJS, it's hard to know whether ovarian tumors in 53 women is a significant finding.

Most of what is written about sex cord tumors is written by pathologists who experience them through a microscope. Because they are microscopic- usually under 3 cm, they are difficult to diagnose by imaging techniques or manual exam. Most are diagnosed after oophorectomy for other problems. This isn't to say that people with sex cord tumors were asymptomatic, just that the sex cord tumors weren't discovered until after surgery. Many females with PJS present with other ovarian tumors & are found to have ovarian sex cord tumors only on pathological examination.

Since you can't look at your intact ovaries through a microscope, it might be worthwhile to know the signs & symptoms of SCTAT and genital tract tumors. If you have concerns based on your experience and what you read here, you might take this information to your doctor or gynecologist. I saw a gynecological oncologist who specialized in



hereditary cancer syndromes who knew nothing about PJS. I gave her some articles and a list of gynecological tumors which led to a discussion and plan for my health management. Unfortunately, doctors are often too busy to learn about PJS. If they are open to it, we can supply articles and information that could improve our health care.

- In boys- gynecomastia (swelling of the breasts), testicular enlargement (sometimes), growth spurts, abnormal hormone levels and advanced bone age.
- In girls- sexual precocity and ovarian mass.
- In women- menstrual irregularity- heavy periods, bleeding between periods, loss of periods, vaginal spotting, bleeding after menopause; ovarian mass; abdominal pain & tenderness; mucous discharge; uterine prolapse; hirsutism and sterility.

If you are interested in SCTAT in females, I suggest getting a copy of *Ovarian Sex Cord Tumor with Annular Tubules: Review of 74 Cases Including 27 with Peutz-Jeghers Syndrome and Four with Adenoma Malignum of the Cervix*. published in **Cancer** 50:1834-1402, 1982. It is a basic text on SCTAT and does us the service of breaking out the PJS cases from the other 47 cases. Their table 1 lists all 27 PJS patients with their ages, signs & symptoms, characteristics of SCTAT, endometrial findings, other findings and follow-up.

Of their 27 PJS patients with SCTAT, 11 had had menstrual irregularity. Nine had pelvic or abdominal masses caused by adnexal (ovarian) tumors of other types. Four had abdominal or pelvic pain or tenderness. And four had Adenoma malignum (a rare cervical cancer associated with PJS).

PJS experts have suggested regular pelvic exams and pelvic ultrasound for females and testicular exams for boys. Search our archives for the topic "screening guidelines". I am uncertain what follow-up or treatment is suggested for the microscopic tumors, but surgery seems indicated for the larger associated ovarian tumors. There are numerous types of ovarian tumors that have been reported in women with PJS. If you are interested in a list, let me know.

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