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Article begins on next page

Pseudopapillary Granulosa Cell Tumor-a case of this rare subtype.

Debra Heller, MD*,**, Andrew Haddad MD**, Bernadette Cracchiolo, MD, MPH**

From the Departments of Pathology & Laboratory Medicine*, and Obstetrics, Gynecology, and Women's
Health**, Rutgers-New Jersey Medical School, Newark, NJ

Address Correspondence to:

Debra S. Heller, MD

Dept of Pathology-UH/E158

Rutgers-New Jersey Medical School

185 South Orange Ave

Newark, NJ, 07103

Tel 973-972-0751

Fax 973-972-5724

hellerds@njms.rutgers.edu

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Precis:

A rare case of pseudopapillary granulosa cell tumor is presented. Knowledge of this subtype will allow inclusion in a differential diagnosis.

Keywords: ovarian neoplasms; granulosa cell tumor; diagnosis, differential

Abstract:

Background: The pseudopapillary pattern of granulosa cell tumor is rare.

Case: We describe a the case of a 35-year-old woman who presented with an initial diagnosis of papillary serous cystadenocarcinoma

Results: Evaluation, including immunohistochemistry, led to the diagnosis of pseudopapillary granulosa cell tumor.

Conclusion: The pseudopapillary pattern of granulosa cell tumor is rare, and must be suspected in order to utilize appropriate immunohistochemistry and reach the correct diagnosis. Inhibin positivity is particularly helpful.

Introduction

Granulosa cell tumors can show a variety of histologic patterns. One of the rarest is a pseudopapillary pattern, with only a few reported cases(1). The differential diagnosis of a papillary pattern in ovarian tumors is large(2). It is important to be familiar with this variant of granulosa cell tumor, in order to suspect it and institute appropriate therapy. We present a case initially diagnosed as papillary serous cystadenocarcinoma.

Case

A 35 year old female patient was referred to gynecology oncology for a diagnosis of metastatic serous papillary carcinoma to the rectum. Past history was significant for right oophorectomy for granulosa cell tumor ten years prior. She had been initially treated elsewhere for the current problem, and computerized axial tomography had shown a 3.7 x 3.2 x 3.4 cm right adnexal lesion. Magnetic resonance imaging six weeks later showed a tubular complex right adnexal mass that was now 8x5x6 cm, suspicious for fallopian tube cancer.

The patient then underwent exploratory laparotomy and excision of metastatic tumor to the rectum. Intraoperatively, two masses measuring one and four centimeters in largest dimensions respectively were found at the level of the anterior wall of the rectum. These were excised as were lesions described as bowel cyst, omental cyst, pelvic cyst, right sidewall cyst and colon cyst. The initial diagnoses were read as papillary serous cystadenocarcinoma.

On presentation upon referral to our institution, she complained of right lower quadrant pain, bloating, and low back pain, as well as a 43 pound weight loss. Review of the outside slides showed recurrent granulosa cell tumor, pseudopapillary type (figures 1,2,3). The tumor was strongly positive for inhibin, WT-1, and focally for calretinin. She then underwent a positron emission tomography (PET) scan showing only a 1.2cm right obturator lymph node with mild uptake. She underwent a hysterectomy with left salpingo-oophorectomy, bilateral pelvic lymph node sampling and low right periaortic lymph node sampling, omentectomy, and debulking of tumor from the left pelvic side wall. Histopathology revealed negative lymph nodes with granulosa cell tumor implants on the left fallopian tube, and rectosigmoid. She had a mediport placed postoperatively for chemotherapy.

Comment:

The pseudopapillary variant of granulosa cell tumor is rare, with only one report of 14 ovarian cases, arising in both juvenile and adult granulosa cell tumors(1). Most of the cases in this series were received in consultation. The pseudopapillae are degenerative, rather than representing true papillae with fibrovascular cores(1). The differential diagnoses raised in Irving's series(1) included transitional cell carcinoma, and a retiform Sertoli-Leydig cell tumor. This rare pattern has also been reported in the uncommon granulosa cell tumors of testis as well(3). Our case was initially called a papillary serous cystadenocarcinoma, in spite of the prior history of granulosa cell tumor, and young age of the patient. Granulosa cell tumors are low grade malignancies that can recur many years later, even after ten years, as in this case. A helpful feature of pseudopapillary granulosa cell tumors is the presence of more characteristic

granulosa areas in the tumor, which were present in this case. Inhibin and calretinin immunostaining are useful confirmatory studies.

The differential diagnosis of papillary granulosa cell tumor includes papillary serous cystadenocarcinoma, retiform Sertoli-Leydig cell tumor and transitional cell carcinoma of the ovary. Papillary serous carcinoma would demonstrate a significantly greater degree of nuclear atypia and would be negative for inhibin and calretinin, and strongly positive for epithelial membrane antigen. Retiform Sertoli-Leydig cell tumor would have a similar immunoprofile to granulosa cell tumor. Helpful distinguishing features are the younger age of patients with Sertoli-Leydig cell tumors, and well as the frequently more typical areas of Sertoli-Leydig pattern in areas of the tumor(4). Transitional cell carcinoma of the ovary is rare, shows more nuclear atypia than granulosa cell tumor, would be expected to be negative for inhibin and calretinin, and stain for CA-125 and estrogen receptor(5).

In summary, it is important to be aware of the possibility of a pseudopapillary pattern of granulosa cell tumor, as the prognosis and treatment can differ significantly from other lesions in the differential. Consideration of a granulosa cell tumor should be given even when there is a remote history.

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Legend:

Figure 1-Granulosa cell tumor showing a pseudopapillary pattern.

Figure 2-Higher power of the lesion shows fairly uniform nuclei with finely dispersed chromatin.

Nuclear grooves were not prominent.

Figure 3-The lesion showed strong inhibin staining.