A 67-Year-Old Woman With Abdominal Distention, Vaginal Bleeding, and Elevated CA 125 Level

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A 67-year-old, gravida 2, para 2, African American woman presented to our institution in November 2004 with vaginal bleeding and worsening abdominal distention for 6 months. The vaginal bleeding was scanty, painless, and intermittent. She had a history of long-term hypertension, hypercholesterolemia, and bilateral osteoarthritis of knees and hands. Her medications were limited to antihypertensives and multivitamins. She was not on hormonal replacement therapy. On physical examination, auscultation of the heart and lungs were unremarkable. Abdominal examination showed a large, mostly solid mass. The mass was palpated in all abdominal quadrants. Its exact size could not be determined on physical examination. Hepatosplenomegaly and lymphadenopathy were not found. Results of the complete blood count, liver enzymes, and renal function tests were all within the reference range. The fecal occult blood test was negative for blood. On admission, abdominopelvic sonography and contrast-enhanced computed tomography showed a large, well-defined, solid, and cystic mass in the pelvic cavity extending to the upper abdomen (Figure 1, arrows pointing to the mass). The mass was found to be involving the left ovary. No retroperitoneal adenopathy was present. Serum CA 125 level was 131.5 U/mL (reference range, <36 U/mL). Subsequently, the patient underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy. Para-aortic, pelvic, and iliac lymph nodes were sampled. Gross examination revealed a large, yellow to brown, predominantly solid mass with multiple hemorrhagic cysts. The mass weighed 6330 g, measured 35 × 30 × 15 cm, and was found to be originating from and replacing the left ovary. Microscopically, the tumor was predominantly composed of hollow to solid, well-defined tubules separated by stroma (Figure 2, A). The tubules were lined with cuboidal to columnar cells with abundant pale to slightly eosinophilic cytoplasm and with bland nuclei showing no mitotic activity (Figure 2, B). In addition to the predominant round tubules, cells were also arranged in cords and aggregates (Figure 3, A) with oval to round bland nuclei (Figure 3, B). Focal areas showing sheets of cells (Figure 4, A) with spindle-shaped nuclei (Figure 4, B) were also seen within the tumor.

What is your diagnosis?
Pathologic Diagnosis: Pure Sertoli Cell Tumor of the Ovary With Differentiation Varying From Well-Differentiated Tubules, to Intermediate Foci, to Sarcomatoid Spindle Cell Areas

Abstract

Pure Sertoli cell tumor of the ovary is a rare sex cord tumor. We describe a case of a pure Sertoli cell tumor in a 67-year-old woman. The tumor exhibited the typical macroscopic and microscopic features. Interestingly, the Sertoli cells were arranged in several microscopic patterns: well-defined tubules, cords and aggregates, and sarcomatoid pattern. The tumor was estrogenic as evidenced by endometrial hyperplasia, which explained the vaginal bleeding. The preoperative CA 125 level was elevated (131.5 U/mL; reference range, <36 U/mL) and normalized postoperatively. Similar to ovarian tumors of epithelial origin, CA 125 level can be elevated in sex cord tumors including pure Sertoli cell tumors. Our case supports the concept of using the measurement of CA 125 level as a predictor of prognosis, curability, and clinical behavior of ovarian tumors as well as a confirmatory tool for posttreatment disease-free state.

Pure Sertoli cell tumor (SCT) is a sex cord tumor as designated by the World Health Organization. This tumor is rare. It may be found in patients of any age, with an average age of 30 years. Sertoli cell tumor may be non-functioning, estrogenic, or occasionally androgenic. Estrogenic effects have been present in two thirds of the reported cases. All the SCTs reported to date have been unilateral. Sertoli cell tumors average 9 cm in diameter and typically form lobulated, solid, yellow, or brown masses. The emergence of such male-oriented cells as neoplastic cord tumors including pure SCTs has been noted in the third week after surgery indicating complete removal of the tumor. Follow-up of the patient using serial CA 125 level changes. Our case also supports the concepts of tumor progression or recurrence without the need for exploratory laparotomy.

In summary, we present a rare sex cord tumor of the ovary. Our case represents a pure SCT of the ovary. The tumor exhibited the typical macroscopic and microscopic features. Pure SCT has several microscopic patterns that can coexist in the same tumor. Elevation of CA 125 level in our case is of interest. Even benign ovarian tumors can present with elevation of CA 125 level; complete removal of the tumor offers a very good prognosis that can be reflected by the CA 125 level changes. Our case also supports the concepts of well-differentiated tubules, intermediate foci, and sarcomatoid spindle cell areas.
that the preoperative values of CA 125 usually have prognostic significance and that changes in the concentrations of CA 125 parallel the course of the disease and treatment response.

References


