

Lipid-Rich Pure Sertoli Cell Ovarian Tumor

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Citation

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Abstract

Sertoli-Leydig tumors (SCT) account for less than 0.5 percent of ovarian tumors, belonging to the category of ovarian tumors derived from specialized gonadal stroma. Average age of onset is the second or third decade of life, but women of all ages can be affected (1). These tumors can be further divided based on the amount of Sertoli or Leydig cells in diverse levels of differentiation. Pure Sertoli cell tumors are rare, lack the Leydig component, and do not contain the immature neoplastic stroma found in Sertoli-Leydig tumors (2). The lipid rich variant is even rarer. We present a case of a lipid rich pure Sertoli cell tumor in an 83 year old woman.

REPORT OF A CASE

An 83-year-old Caucasian female, presented to the emergency department for abdominal pain and vaginal bleeding. Abdominopelvic sonography and tomography showed a solid mass involving the right ovary. Neither uterine enlargement nor endometrial thickening was observed. Laboratory test revealed elevation of tumor markers CA 125 (51,3 U/ml, reference range <30 U/ml) and CA 19-9 (35 U/ml, reference range < 31 U/ml). The levels of hCG, AFP in the serum were normal. She had no evidence of androgen excess or virilization. Testosterone estimations were not carried out. A laparoscopy was performed with findings of an encapsulated, mostly solid mass of the right ovary, so hysterectomy, and bilateral salpingo-oophorectomy was performed. The specimen was sent to the Pathology Department. Gross examination revealed a predominantly solid but soft yellow tumor, measuring 5 x 4 x 3 cm and weighing 300 g (Figure 1). Microscopically, the tumor was predominately composed of solid nests of cells, arranged in vague nodules separated by thin fibrous bands. The nest were composed of cuboidal to columnar cells with striking clear cytoplasm due to lipid as proved by positive fat stain (oil-red-or) (Figure 2A, 2B). Round to oval regular nuclei with inconspicuous nucleoli and no appreciable cytological atypia were seen. No Leydig cells were found and the mitotic rate was very low. Immunohistochemistry was performed showing strong positivity to β -inhibin (Serotech, Oxford, UK, dilution 1:12) (Figure 3A,3B), Melan A (Dako, Carpinteria, CA, dilution 1:60) (Figure 5), Calretinin (Zymed, South San Francisco, CA, dilution neat)

and AE1/AE3 (Dako, Carpinteria, CA, dilution 1:30).

Figure 1

Figure 1: Yellow-white fragile solid tumor with lobulated appearance.

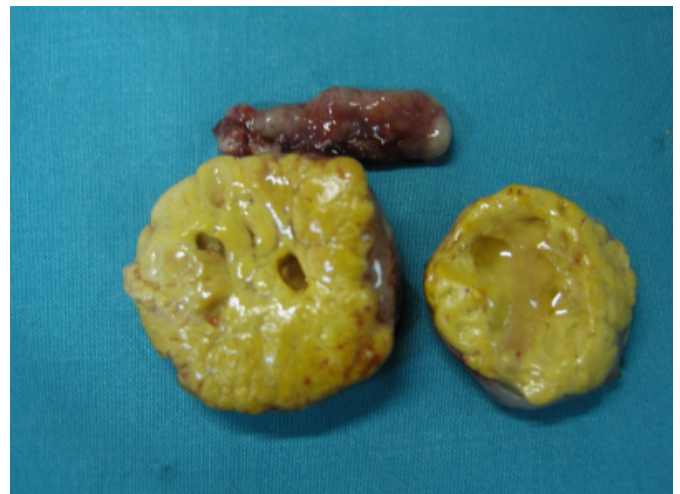


Figure 2

Figure 2A and 2B: Anastomosing solid tubules separated by thin fibrous bands. Tumor cells have clear cytoplasm due to lipid as proved by a positive fat stain (2A Hematoxylin and eosin, Original magnification x20). (2B Oil red-or, original magnification x40).

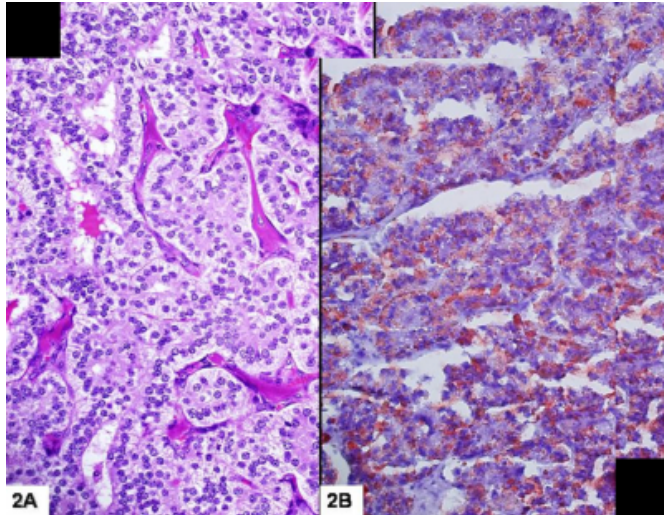
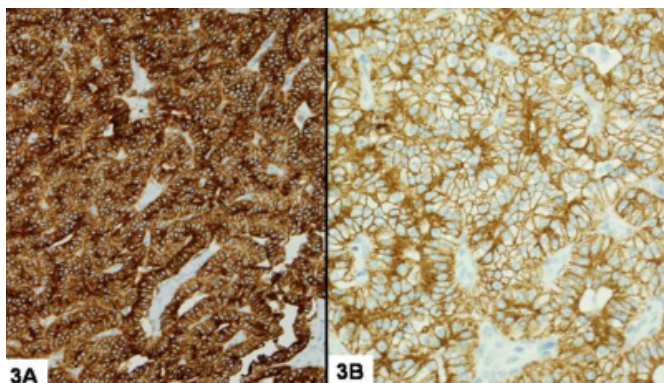


Figure 3

Figure 3A and 3B: Diffuse positivity for β -inhibin (3A) and Melan A (3B). (Peroxidase, original magnification, 20x)



COMMENT

Pure Sertoli cell tumors are rare ovarian tumors of the sex-cord stromal cell origin as designated by the World Health Organization [34]. They account for about 0.5 % of all ovarian neoplasm [15]. They are almost always unilateral, occurring in women 30 year old or younger. Fewer than 10% of the patients are over 50 years of age. About 30-40% of patients show virilization [6]. There have been four large histopathological studies of Sertoli-stromal cell tumors totalling more than 350 cases [6,78910]. According to these reports, the gross appearance reflects varied components: solid, solid/cystic (the most common) or totally cystic. Microscopically, they range from well to poorly

differentiated. SCT are classified based on histological pattern: simple tubular, complex tubular and lipid rich type. By definition pure Sertoli cell tumors lack Leydig cell in the stroma [3]. However, heterologous elements such as mucinous glands, bone, skeletal muscle and cartilage may be present in some tumors [34]. Immunohistochemical staining of SCT typically shows reactivity for cytokeratin and β -inhibin. Positivity for melan A has also been described as a useful marker [11]. The tumor is typically negative for epithelial membrane antigen, carcinoembryonic antigen, placental alkaline phosphatase and S100 [3]. Differential diagnosis of SCT includes mucinous tumors, low grade endometrioid carcinoma, carcinosarcoma when heterologous elements are present, tubular Krukenberg tumor, tubular carcinoid and ovarian tumors of probable Wolffian origin [31213].

The prognosis of SCT is usually good and correlates with the stage and degree of differentiation of the tumor [3]. Very rarely SCTs show malignant histological features including high mitotic rate, necrosis, marked cytological and nuclear atypia, and lymphovascular invasion. Adjuvant therapy is considered based on the histological classification and staging of the tumor. Poorly differentiated or metastatic tumors have noted to have a poor prognosis without chemotherapy [10].

In the current case, SCT was confined to the right ovary, International Federation of Gynecology and Obstetrics, stage IA. The tumor showed the typical macroscopic and microscopic features discussed earlier, without any of the features of worse prognosis, thus a benign course would be expected. As published before, serum CA 125 level was elevated, decreasing its level after surgery and becoming normal four week after surgery. CA 125 could be used in SCT as a marker of recurrence [14].

In summary, we present a rare well differentiated lipid rich pure sex cord tumor in a woman on her eighties, with the typical macro and microscopic features and elevation of CA 125.

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