Case Reports

Sertoli cell tumor: a rare case in an elderly patient

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Summary

Sertoli-Leydig cell tumors constitute < 1% of ovarian tumors, mostly in young women with virilization; however, not all present endocrine manifestations.

A 72-year-old female presented with an abdominal mass and no signs of virilization. Total abdominal hysterectomy with bilateral salpingo-oophorectomy, omentectomy and selective pelvic lymphadenectomy was performed. The pathologic diagnosis was poorly-differentiated sex cord-stromal tumor with Sertoli cells. No adjuvant chemotherapy or radiation was administered. At 12-month follow-up the patient showed no evidence of disease.

Key words: Sertoli-Leydig cell tumors; Elderly; Ovarian cancer.

Introduction

Ovarian cells of sex-cord origin manifest a capacity for testicular differentiation in neoplasms. Because less-differentiated forms may mimic the development of the testes, the terms androblastoma and arrhenoblastoma have erroneously been used as synonyms for Sertoli-Leydig cell tumors (SLCT): some SLCT have no endocrine manifestation while others may be accompanied by an estrogenic syndrome. SLCT are most often encountered in young women, with or without virilization [2-4].

Case Report

A 72-year-old female presented with an abdominal mass. The patient was obese with normal breasts and sparse axillary and pubic hair. There was no evidence of virilization. Abdominal ultrasound demonstrated the presence of a non-homogeneous, well-vascularized right adnexal mass 10 cm in diameter with an irregular profile, later confirmed by CT.

Total abdominal hysterectomy with bilateral salpingo-oophorectomy, omentectomy, selective pelvic lymphadenectomy, peritoneal washing, and random biopsies were performed. The histopathological findings revealed poorly differentiated malignant sex cord-stromal tumor with Sertoli cells. The uterus, contralateral ovary and lymph nodes were free of disease. Peritoneal washing was negative. The postoperative course was uneventful. The patient continued to show no evidence of disease 13 months later.

Results

The case was a rare postmenopausal Sertoli-Leydig cell tumor. Cases reported in elderly patients describe a sudden onset of severe, rapid virilization associated with a tumor source of androgens. The patient showed no sign of virilization.

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SLCT are classified into five histological types with different prognoses [5]. In immunocytochemical characterization of SLCT the Sertoli and Leydig cells stain positive for testosterone and estradiol. The areas with Sertoli cells are positive for keratins and vimentin, and negative for epithelial membrane antigen (EMA), placenta-like alkaline phosphatase (PLAP), carcinoembryonic antigen (CEA), CA 19.9, CA 125 and S-100 protein. SLCT with hepatocytic differentiation are positive for alpha-fetoprotein (AFP). Our poorly differentiated SLCT stained positive for AFP, PLAP, CD99, MFN 116, progesterone receptor, vimentin and negative for CEA, β-HCG, estrogen receptor, S-100.

Over 90% of SLCT cases are unilateral (usually the left ovary), located in the ovarian medulla, sometimes multifocal [6]. Our case presented a right ovarian tumor, 10 cm in diameter. SLCT are solid tumors with dimensions that correlate with the degree of differentiation: well-differentiated tumors are smaller than 0.5 cm, while poorly differentiated tumors can reach up to 10-15 cm.

Relapse after surgical treatment of SLCT has been reported in 33% of cases and is more common in poorly differentiated tumors, even in Stage IA [7]. Metastases have been observed in the omentum, abdominal lymph nodes and liver [8]. No metastases were found in this case, which was classified as Stage IA.

Because SLCT are rare, their management is controversial. The debate is over the type of surgery, radical or not, and whether radiation or chemotherapy is necessary. Treatment depends on the histological characteristics as well as patient age. Unilateral salpingo-oophorectomy is the treatment of choice for young women in Stage I, while hysterectomy with bilateral oophorectomy is appropriate for older women. There is general agreement that patients with metastatic tumor (Stage II-IV or recurrence) or with poor prognostic factors should receive therapeutic treatment. Prognostic factors other than stage appear to be histologic differentiation, mitotic index, the presence of heterologous

Table 1. — *Recent cases of SLCT.*

Authors	Country	Year	N° Cases	Age	Secreting Yes/No	Stage	Grade	Surgery	CT	RT	Result
Gershenson [9] *et al.*	USA	1996	2	22	NS	Ia	3	Before and after CT for relapse of disease (cytoreductive surgery)	BEP+ paclitax+ DDP	no	31 mo: NED
				14	NS	Ia	3	Before CT	BEP	no	28 mo: NED
Lantzsch [7] *et al.*	Germany	2001	1	11	Yes: A/E/P	I	2	Left salpingo-oophorectomy	no	yes	6 mo: NED
Gheorghisan [10] *et al.*	Romania	2003	1	69	Yes: A	NS	1	Hysterectomy, bilateral oophorectomy	no	no	3.5 yrs: NED
Appetecchia [11] *et al.*	Italy	2004	1	23	Yes: A/E	Ia	1	Excision of neoplasm	no	no	24 mo: NED
Lara-Torre [12] *et al.*	USA	2004	1	19	Yes: A	IIc	3	Left salpingo-oophorectomy, endometrial biopsy, lymph node sampling	BEP	no	10 mo: NED
Tampakoudis [13] *et al.*	Greece	2004	1	71	No	I	1	Hysterectomy, bilateral oophorectomy, lymph node sampling excision vaginal lesion	no	no	26 mo: NED
Nicoletto *et al.*	Italy	2006	1	72	No	I	3	Hysterectomy, bilateral oophorectomy, lymph node sampling	no	no	13 mo: NED

A = androgen; E = estrogen; P = progesterone; NS = not specified; NED = no evidence of disease; CT = chemotherapy; BEP = bleomycin +etoposide+cisplatin; paclitax = paclitaxel; DDP = cisplatin; RT = radiation therapy.

elements and tumor capsule rupture. Gershenson *et al.* treated two poorly differentiated Stage IA SLCT with surgery and chemotherapy. After completion of four cycles of bleomycin/etoposide/cisplatin, one case was disease-free at 28 months and one relapsed 17 months after diagnosis. She was treated with secondary cytoreductive surgery followed by six cycles of chemotherapy with paclitaxel/cisplatin and was disease-free 31 months after diagnosis [9] (Table 1).

It is important to continue to collect data on this rare form of neoplasm in order to better understand the most appropriate treatment after surgery: adjuvant or surveillance.

References

[1] Ergete W., Kiros K.: "An ovarian Sertoli-Leydig cell tumour in a 40 year old Ethiopian woman: case report". *Ethiop. Med. J.*, 2000, *38*, 185.

[2] Zaloudek C., Norris H.J.: "Sertoli-Leydig tumors of the ovary". *Am. J. Pathol.*, 1984, 8, 405.

[3] Young R.H., Scully R.E.: "Ovarian Sertoli-Leydig cell tumours". *Am. J. Surg. Pathol.*, 1985, *9*, 543.

[4] Britt K.L., Findlay J.K.: "Regulation of the phenotype of ovarian somatic cells by estrogen". *Mol. Cel. Endocrinol.*, 2003, *202*, 11.

[5] Young R.H.: "Sertoli-Leydig cell tumors of the ovary: Review with emphasis on historical aspects and unusual variants". *Int. J. Gynecol. Pathol.*, 1993, *12*, 141.

[6] Latthe P., Shafi M.I., Rollason T.P.: "Recurrence of Sertoli-Leydig cell tumor in controlateral ovary. Case report and review of literature". *Eur. J. Gynaecol. Oncol.*, 2000, *21*, 62.

[7] Lantzsch T., Stoerer S., Lawrenz K.: "Sertoli-Leydig cell tumor". *Arch. Gynecol. Obstet.*, 2001, 206.

[8] Campisi P., Cheski P.: "Metastatic Sertoli-Leydig cell ovarian cancer manifested as a frontal sinus mass". *J. Otolaryngol.*, 1998, *27*, 361.

[9] Gershenson D.M., Morris M., Burke T.W.: "Treatment of poor-prognosis sex cord-stromal tumors of the ovary with the combination of bleomycin, etoposide and cisplatin". *Obstet. Gynecol.*, 1996, *87*, 527.

[10] Gheorghisan-Galateanu A., Fica S., Terzea D.C.: "Sertoli-Leydig cell tumor - a rare androgen secreting ovarian tumor in post-menopausal women". *J. Cell Mol. Med.*, 2003, *7*, 461.

[11] Appetecchia M., Cela V., Bernardi F., Burelli A., Cionini R., Pucci E.: "Sertoli-Leydig cell androgens-estrogens secreting tumor of the ovary: ultra-conservative surgery". *Eur. J. Obstet. Gynecol. Reprod. Biol.*, 2004, *116*, 113.

[12] Lara-Torre E., Dietrich J.E., Kaplan A., Lopez H., Jaffee I.: "A case of poorly differentiated Sertoli-Leydig tumor of the ovary". *J. Pediatr. Adolesc. Gynecol.*, 2004, *17*, 49.

[13] Tampakoudis P., Zafrakas M., Kostopoulou E.: "Ovarian Sertoli-Leydig cell tumor with coexisting vaginal angiomyxoma: case report and review of the literature". *Eur. J. Gynecol. Oncol.*, 2004, *25*, 116.

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