Ovarian Sertoli-Leydig cell tumor with coexisting vaginal angiomyxoma: Case report and review of the literature

P. Tampakoudis[1], M. Zafrakas[1], E. Kostopoulou[2], K. Dragoumis[1], T. Tsalikis[1], J. Bontis[1]

[1]1st Department of Obstetrics and Gynaecology, Aristotle University of Thessaloniki, Hippokrateio General Hospital
[2]Department of Pathology, Hippokrateio General Hospital, Thessaloniki (Greece)

Summary

An extremely rare case of a postmenopausal patient with an ovarian Sertoli-Leydig cell tumour and a coexistent vaginal angiomyxoma is reported. A 71-year-old patient was admitted complaining of abdominal distension. A thorough diagnostic evaluation revealed a large tumour of the right ovary, and an oval-shaped greyish-white polypoid vaginal lesion. Total hysterectomy with bilateral salpingo-oophorectomy and lymph node sampling was performed, followed by excision of the vaginal lesion. Histological examination showed a Sertoli-Leydig cell tumour of the right ovary, and a vaginal angiomyxoma. Twenty-six months after the operation the patient is well with no signs of recurrence. To the best of our knowledge, no case of coexistence of an ovarian Sertoli-Leydig cell tumour with a myxoma has been previously reported.

Key words: Sertoli-Leydig cell tumour; Vaginal angiomyxoma; Carney complex.

Introduction

Sex cord-stromal tumours of the ovary account for approximately 5% to 8% of all ovarian malignancies. They are usually composed of various combinations of elements, including "female" type cells, i.e. granulosa and theca cells, and less frequently "male" type cells, i.e. Sertoli and Leydig cells [1]. Sertoli-Leydig cell tumours are extremely rare; it is estimated that they account for less than 0.2% to 0.5 % of all ovarian malignancies. It has been reported that 75% of these tumours occur in women younger than 40 years of age, and that the average age of patients is 25 years [2-5].

Case Report

A 71-year-old patient, gravida 2, para 2, was admitted complaining of abdominal distension, and minor vaginal bleeding four days before admission, which resolved spontaneously. The patient's last menstrual period was 24 years earlier; she suffered from chronic obstructive pulmonary disease, essential hypertension, and had a cerebral vascular accident one year before admission. Pelvic examination revealed an oval-shaped greyish-white exophytic lesion at the upper third of the left vaginal wall, with a diameter of approximately 2.5 cm, and a large, painless, solid tumour extending into the abdominal cavity could be easily palpated. A large multilobular pelvic mass with both solid and cystic components was visualized on ultrasound and CT.

The patient underwent exploratory laparotomy. A mixed solid-cystic mass of the right ovary, measuring approximately 16 cm, was removed together with the right adnexum, and total hysterectomy with left salpingo-oophorectomy and sampling of enlarged lymph nodes around the right iliac artery followed. The operation was completed with excision of the vaginal lesion.

Microscopic examination of peritoneal washings was negative for malignant cells. Microscopic examination of multiple histological sections showed characteristics of a Sertoli-Leydig cell tumour, of intermediate-low differentiation, with multiple mitoses (Figure 1). Immunohistochemical stains for inhibin, CD99, and vimentin were positive. There were weakly positive foci in immunohistochemical stains for cytokeratins, whereas stains for NSE, GFAP, and thyreoglobulin were negative. The right fallopian tube, the uterus, the contralateral adnexum and five iliac nodes did not show any signs of malignancy. In regard to the vaginal lesion, the histological findings suggested the presence of an angiomyxoma (Figure 2).

The patient did well during the immediate postoperative period and is still disease-free 26 months after the initial diagnosis.

Discussion

There are no Sertoli cells in the normal adult ovary. In neoplasia ovarian cells of sex cord origin manifest capacity for testicular differentiation. The Leydig cells, which normally are seen in the hilus, appear in the medullary and cortical stroma in neoplasia [6]. The two main theories concerning the histogenesis of Sertoli-Leydig cell tumours suggest that they arise either from the gonadal mesenchyme of the ovary or from remnants in the hilum [2]. Heterologous elements are present in about 20% of ovarian Sertoli-Leydig cell tumours [2, 6, 7]. The tumour in the present case showed no such heterologous elements

Sertoli-Leydig cell tumours typically produce androgens; clinical virilization is seen in 70% to 85% of patients [2]. The present case differs from the typical presentation of Sertoli-Leydig cell tumours in the following aspects: 1) there were no signs of virilization. 2) the patient was postmenopausal. 3) Sertoli-Leydig cell tumours are most frequently low-grade malignancies, whereas in the present report, the tumour appeared as an intermediate-high grade tumour.

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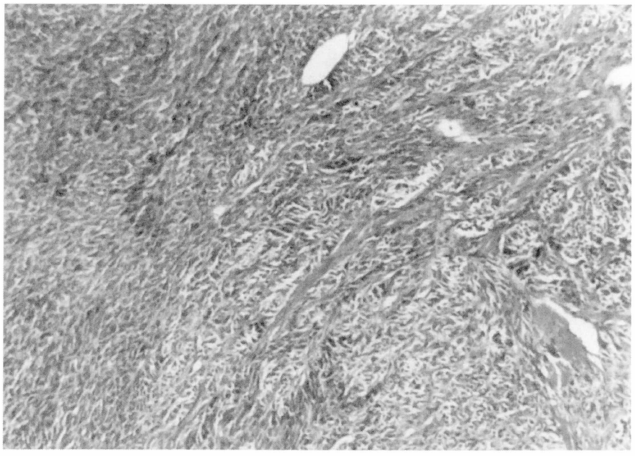

Fig. 2

Figure 1. — Ovarian tumour showing histological characteristics of Sertoli-Leydig cell tumour of intermediate-poor differentiation (HE, original magnification x 200).
Figure 2. — Vaginal lesion with characteristics of angiomyxoma (HE, original magnification x 100).

In postmenopausal patients, as in the present case, hysterectomy and bilateral salpingo-oophorectomy are appropriate. In premenopausal women, the usual treatment is unilateral salpingo-oophorectomy and clinical evaluation of the contralateral ovary, since these tumours very rarely are bilateral (< 1%). Tumour stage higher than I requires more aggressive surgery and adjuvant therapy [8]. Although the best chemotherapy for Sertoli-Leydig cell tumours is not known, a platinum-based one is generally favoured [6]. Due to early disease stage, and patient age and general medical status no postoperative treatment followed in this case.

Angiomyxomas are rare, benign neoplasms derived from connective tissue with large numbers of vascular structures. They consist chiefly of polyhedral and stellate cells, loosely embedded in a soft mucoid matrix, thereby resembling primitive mesenchymal tissue. In the female genital system less than 100 cases of angiomyxomas have been reported. In most of these reports "superficial" and "aggressive" angiomyxomas of the vulva and perineum have been described, while only a few cases of "aggressive" angiomyxomas of the vagina have been reported. These lesions never metastasize, but late local recurrence is common making long-term follow-up imperative [9-11].

Coexistence of other tumours with ovarian Sertoli-Leydig cell tumours is rarely reported. Cervical sarcoma botryoides has been reported to occur in three cases prior to the discovery of an ovarian Sertoli-Leydig cell tumour [3, 12]. Other ovarian tumours that have been reported to occur in patients with Sertoli-Leydig cell tumours include clear cell adenocarcinoma, serous cystadenoma, mature and immature teratoma [2, 3]. A history of multinodular goiter or thyroid tumours has also been reported in a few cases [3, 13].

Coexistence of testicular Sertoli-Leydig cell tumours and myxomas has been previously described in men in the context of a multiple neoplasia and lentigosis syndrome referred to as the Carney complex. The Carney complex is an autosomal dominant, genetically heterogeneous syndrome characterized by: spotty skin pigmentation, myxomas, endocrine hyperactivity, and schwannomas [14-16]. In male patients, testicular Sertoli-Leydig cell tumours are commonly seen, whereas in a recent study in women with Carney Complex, in which 18 patients were studied prospectively, and 178 retrospectively, no case of ovarian Sertoli-Leydig cell tumour was found [17].

In the present case, the coexistence of an ovarian Sertoli-Leydig cell tumour with an angiomyxoma led to a detailed examination of the patient during follow-up, with special attention for features of the Carney complex. The patient indeed had spotty-pigmentation of the skin in the dorsum and upper extremities; CT scan of the abdomen, bilateral mammography and echocardiography, all done 26 months after the initial diagnosis, were unremarkable. To the best of our knowledge no case of Carney complex involving an ovarian Sertoli-Leydig cell tumour and no case of coexistence of an ovarian Sertoli-Leydig cell tumour with a myxoma has been previously reported. Close and careful follow-up with special attention for other features of the Carney complex is the management plan. Nevertheless, coexistence of two different neoplasms in the same patient dictates per se close and careful patient follow-up.

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Address reprint requests to:
M. ZAFRAKAS, M.D.
Evzonon 21
54640 Thessaloniki (Greece)