Ovarian Sex Cord Tumor With Annular Tubules

Clinicopathologic Report of Two Benign and One Malignant Cases With Long Follow-Ups

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Summary. The clinicopathologic features of three new cases of ovarian sex cord tumors with annular tubules are presented, thereby increasing to 23 the number of the published cases in the world literature. These three observations, along with another one which was previously published, were found in the files of the Institute of Pathology of the University of Lausanne from 1939 to 1978. Forty-seven granulosa cell tumors and eight Sertoli and/or Leydig cell tumors of the ovary were found during the same 40-year period. The patients were 48, 64 and 71 years of age. No sign of the Peutz-Jeghers syndrome was noticed in the three patients. All three tumors caused metrorrhagias as a cardinal sign. They were bulky, unilateral and were formed by solid tissue with cystic spaces. Histologically, the most characteristic pattern consisted of simple and complex tubular structures as described by Scully in 1970. Two patients, in which the mitotic indexes of the tumors were lower than 5 mitoses per 10 HPF, died without evidence of a recurrence 36 and 37 years after surgical ablation of the tumor. The third patient, whose neoplasm featured fewer well differentiated tubular structures than the two previous ones and had a mitotic index of over 70 mitoses per 10 HPF, died from massive abdominal recurrence after 5 years and 5 months.

Key words: Ovarian neoplasms – Pathology.

The ovarian sex cord tumor with annular tubules (SCTAT) was described by Scully in 1970; at the same time, he recognized its frequent association with the Peutz-Jeghers syndrome. Since then, only a few cases have been reported which were or were not combined with this syndrome. Little is known, however, of its evolution as no long follow-up studies have been reported.

The purpose of this paper is to evaluate the frequency of SCTAT with regard to other ovarian neoplasms of the sex cord mesenchyme and to describe 3 new cases with extended follow-ups. One of these behaved in a malignant fashion and caused the death of the patient.
Materials and Methods

Four cases of SCTAT were found during a review of the ovarian tumors in the files of the Institute of Pathology of the University of Lausanne from 1939 up to 1978. One of them, which was associated with a Peutz-Jeghers syndrome, was previously published (Gloor, 1978) and it is not included in the present series. For the study of the three other cases which are presented here, the original slides were reexamined when available; new ones were prepared and a catamnestic inquiry was undertaken. The mitoses were counted in three series of 10 high-power fields (HPF-400) and the mitotic indexes expressed the minimal and maximal number of mitoses per 10 HPF in each of the three series. Owing to the retrospective character of this study, neither ultrastructural observations nor hormonal determinations could be performed.

Case Reports

Case 1. Following menometrorrhagias, a 48-year old woman, para 2, underwent a supracervical hysterectomy and a left salpingo-oophorectomy for a bulky and partly cystic tumor of the left ovary. There was a cystic and focally atypical hyperplasia of the endometrium. No manifestation of the Peutz-Jeghers syndrome was noticed. The patient died 36 years later, at 84 years of age, from a cranio-cerebral traumatism without evidence of a recurrence of the ovarian tumor. There was no autopsy.

Case 2. A 64-year old woman, para 4, 12 years post-menopausal, presented metrorrhagias. Dilatation and curettage was negative. A left salpingo-oophorectomy was performed for a bilocular cystic tumor of the left ovary. The tumor was thick-walled, partly calcified and was estimated at about 15 cm in diameter. Its pedicle was twisted. No stigmate of the Peutz-Jeghers syndrome was observed. Thirty-seven years later, at 101 years of age, the patient died with signs of cardiac failure without evidence of a recurrence of the ovarian tumor. There was no autopsy.

Case 3. A 65-year old woman, para 9, underwent a left salpingo-oophorectomy for a serous papillary cystadenofibroma of the left ovary estimated at about 10 cm in diameter. No sign of the Peutz-Jeghers syndrome was remarked. At 71 years of age, after metrorrhagias, a total abdominal hysterectomy with a right salpingo-oophorectomy was performed for a tumor of the right ovary. This tumor was partly solid and partly cystic; it measured 13 cm in its largest dimension. The endometrium was proliferative, the glands showing some mitoses; however, there was no cellular nor architectural atypia. The tubal epithelium was high-cylindrical.

Five years and five months later, the patient was reexamined for a massive abdominal recurrence of the tumor of the right ovary which disturbed intestinal function. During operation, solid tissue with small cystic spaces containing an aqueous liquid was found. It adhered to the pelvic walls and infiltrated the sigmoid and a loop of the small intestine which was angulated. The intestinal loop was resected and a colostomy was performed. Several fragments of the tumor were removed, but the bulk of this infiltrating mass had to be left in place. The patient died on the same day. There was no autopsy.

Results

Except for some nuances, the histological features of the three cases including the abdominal recurrence in case 3 are similar and very characteristic (Fig. 1–3): the tumors consist especially of well circumscribed, rounded, oval or polygonal epithelial nests (Fig. 1a, 2a, 3a) which sometimes coalesce into broad fields. These epithelial nests contain hyaline eosinophilic structures composed of small

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