Int J Clin Exp Pathol 2016;9(10):10770-10773
www.ijcep.com /ISSN:1936-2625/IJCEP0020961

Case Report

Malignant intra-mesosalpinx extra-ovarian sex-cord like tumor: a case report

Yu Gu, Wenbin Xu, Bole Zhuang

Obstetrics and Gynecology Hospital Affiliated to Fudan University, Shanghai, China

Received December 1, 2015; Accepted February 13, 2016; Epub October 1, 2016; Published October 15, 2016

Abstract: Uterine sex cord-like tumor (USCLTs) is a type of very rare cancer. It's originally divided into two subtypes: type I and type II, and it is very similar with real sex cord tumor. A 47-year-old Chinese woman; 7 years after total hysterectomy, pelvic masses had been found for 6 years. Transabdominal bilateral adnexa resection, greater omentum resection and pelvic lymph node dissection were performed simultaneously. In the follow up visit, no trend of tumor recurrence showed, according to all previous gynecologic examinations. Eleven months later, abdominal radical right hemicolectomy was performed because of 'stool characters have changed for several years'. Immunohistochemical staining and hematoxylin and eosin (H&E) staining were performed for pathological study. This type of tumor is not a result of primary ovarian sex cord stromal tumor's metastasizing inside the mesosalpinx, neither Wolffian adnexal tumor, but an extra-ovarian sex-cord like stromal tumor that generated in mesosalpinx, which is reported here for the first time.

Keywords: Gynecological tumor, uterine tumor, ovarian sex-cord like tumor

Case report

Medical history

Female patient, 47 years old, hospitalized in August 13, 2012, because of 'Seven years after total hysterectomy, pelvic masses had been found by medical examination for 6 years'. Usually, soreness of waist, abdominal distension and abdominal pain occur, but no other self-reported discomfort symptoms.

Childbearing history: G4P3, 3 eutocias in her early years, surgical ligation performed in 1990, and abdominouterectomy performed in 2005, because of 'pathological changes of endometrium'.

Gynecological examination: vulva: married type, vagina: unobstructed, the top healed well. Cervix and corpus: absent. Bilateral adnexa: incrassated right adnexa area, no obvious abnormity found on the left. B ultrasonic: right ovary size: 16 × 11 × 8 mm^3, medium-low echo area inside: 47 × 38 × 32 mm^3, color blood flow, funicular; left ovarian: size 22 × 9 × 7 mm^3, CA125, CA199, CEA were all normal.

Admitting diagnosis: the nature of the pelvic mass has yet to be investigated. Five days after admission, transabdominal bilateral adnexa resection, greater omentum resection and pelvic lymph node dissection were performed simultaneously. Intraoperative findings: right ovarian cystically dilated by about 5 × 4 × 3 cm^3, the surface was smooth, and there were no ascites in the pelvic cavity. Intraoperative frozen sections indication: right adnexa had poorly differentiated adenocarcinoma.

Postoperative pathological results

Gross inspection showed that it was right fallopian tube, 5 cm long, 0.5 cm in diameter, without fimbriated extremity or cavity expansion. A nodular mass (size, 2 × 2 cm) was found in the mesosalpinx, which was brittle and had yellow-gray or taupe transections. Right ovary: 2.5 × 2.5 × 1 cm^3, no obvious mass was found. By over checking the gross specimen, we found that the tumor had no tissue relationship with the right ovary.

Pathological diagnosis was the malignant extra-ovarian sex-cord like tumor with rhabdoid differ-

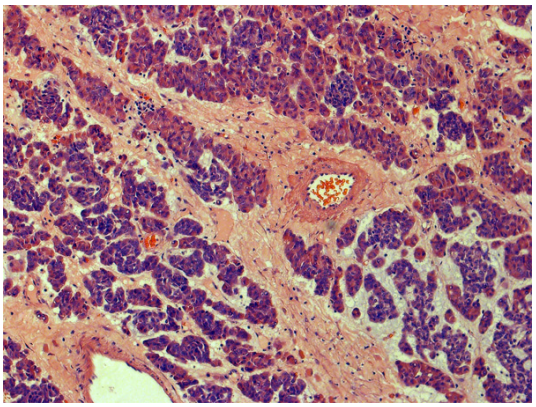

Figure 1. The malignant intra-mesosalpinx extra-ovarian sex-cord like tumor. H&E staining, 200×.

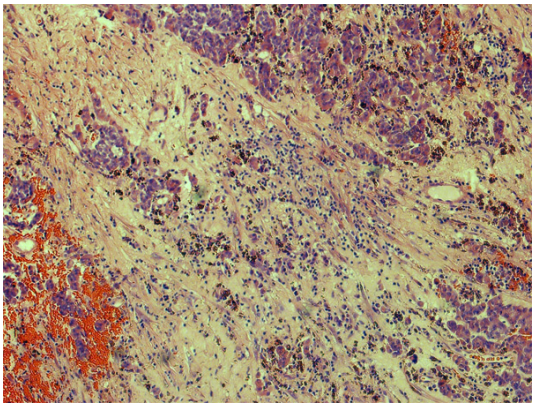

Figure 2. The malignant extra-ovarian sex-cord like tumor with rhabdoid differentiation in right mesosalpinx. H&E staining, 200×.

entiation in right mesosalpinx behaved in a way of invasive growth (**Figures 1**, **2**). However, no obvious pathologic change was found in bilateral ovaries and fallopian tubes. In all of the 6 groups, greater omentum or pelvic lymph node cancer metastasis were not observed.

Immunohistochemistry showed that the tumor cells were AE1/AE3 (modicum, +), CK7 (modicum, +), EMA (-), Vimentin (+++), CA-125 (-), WT-1 (+++), Inhibin-a (+), Calretinin (+++), ER (++), PR (++), P53 (+), Ki-67 (20% +), CgA (-), Syn (-), CD10 (-), Ber-EP4 (-), CK5/6 (-), D240 (-), HBME-1 (-), Myogenin (-), MyoD1 (-), SMA (+/-), Desmin (+), Caldesmon (-), S-100 (-), HMB45 (-).

Postoperative therapy

Postoperative chemotherapy was conducted four times with carboplatin and bleomycin combination.

Follow-up visit

The patient, who showed no trend of tumor recurrence in all previous gynecologic examinations, is in a close gynecological follow-up at present. During the follow-up, the patient was hospitalized in Zhongshan Hospital Affiliated to Fudan University because of the self-report 'stool characters have changed for several years' on July 16, 2013. The 3rd day after admission, abdominal radical right hemicolectomy was performed. The postoperative pathology of right hemicolon showed a protrude-type adenocarcinoma at II differentiation degree, part of which was mucous adenocarcinoma, and the cancerous tissue has infiltrated into full-thickness intestinal wall. No cancer involvement was observed on two cut edges. Besides, we found 14 lymph nodes adjacent to ileum, and all of them showed no cancer metastasis (0/14), and 14 adjacent to colon, all of which neither had no cancer metastasis (0/14), but one cancer nodule and chronic appendicitis existed. In addition, two other lymph nodes were also without cancer metastasis (0/2). A total of four courses of the treatment with combination of oxaliplatin and 5-fluorouracil were postoperatively given. Now the patient has survived healthily for 3 years.

Discussion

Differential diagnosis

Theca cell tumor, fibroma tumor and granular cell tumor are the most common ovarian sex cord stromal tumor, which account for 4.30~6.00% of ovarian tumors [1, 2]. Reproductive cells, derived from the entoderm tissue outside the gonad, can differentiate into both male and female. In the process of its occurrence, migration and development, reproductive cells can mutate to form tumors. These types of tumors secrete estrogen, which causes a series of endocrine disorders. After the resection of the original lesions, they can metastasize or recur, and there are very few ovarian sex cord stromal tumor originated outside of the ovary. Extraovarian sex cord stromal tumors which are presently reported in literature are mainly the granular cell tumors of posterior peritoneum, broad ligament, mesentery, omentum majus, liver, paranephros and other parts [3]. However, the extraovarian sex cord stromal

tumor which occurred in the mesosalpinx hasn't been reported yet. In this case, the possibility that primary ovarian sex cord stromal tumor metastasize inside the mesosalpinx has been ruled out. Therefore, no lesions were found in bilateral ovaries of the patient.

Wolffian adnexal tumor (WAT) is located in broad ligament, mesosalpinx and ovaries, where there are no mesenchymal cells, and it is a relatively rare tumor whose biological behavior is difficult to determine. Clinically, hormone secretion is not generally present in WAT like Sertoli-Leydig cell tumor. The results of immunohistochemical staining were as follows: CK (AE1/AE3) Vim CK19 (+), CK7 EMA CR (+, partly), Ki67 (+, 40%), Inhibin-α CD10 (-) [4]. Nevertheless, the results of immunohistochemical analysis of the patient didn't match with WAT perfectly.

Uterine sex cord-like tumors (USCLTs) is a very rare tumor [5]. It was put forward for the first time by Clement and Scully in 1976 and named as an ovarian sex-cord tumor-like uterine tumor [6]. According to the various ratios of sex cord differentiated composition and endometrial stromal tumors composition, the authors divided it into two subtypes [5, 6]: Type I, mainly composed of endometrial stromal tumor ingredients, and reversible focal accompanied with ovarian sex cord tumor-like epithelioid differentiated composition; easy to relapse. Type II, almost all or most were composed of sex cord-like composition. The tumor perimeter was clear and not easy to relapse, and its prognosis was better than that of type I. In 1988, Clement et al. suggested the type I to be named as endometrial interstitial cell tumor with epithelioid differentiation, and the type II to be named as uterine tumors resembling ovarian sex-cord tumors [7, 8]. Classification of female genital tumor by WHO defined it as uterine sex cord-like tumors (USCLTs), a kind of corpus uteri tumor which was very similar to real sex cord tumor [9]. It was emphasized that only when the tumor had no other compositions like classic endometrial stromal tumors or smooth muscle tumor, could it be diagnosed. In 2005, panhysterectomy was performed on the patient because of endometrial lesions. However, the specific postoperative pathology is unclear. According to the biological characteristics of uterine sex cord-like tumor, we can't exclude for sure that the pelvic tumor has nothing to do with it. It needs to be further confirmed after inquiring the medical history in detail and reviewing the pathological results.

Family history of cancer

There were six siblings in the patient's father generation, three of which had died of cancer. The patient's father underwent gastric carcinoma operation, and died of liver cancer 10 years later. Meanwhile, her father's elder brother died of esophageal cancer; the younger brother suffered from gastric carcinoma, and died of bladder cancer later in the end. The patient's grandfather died of gastric carcinoma. There were 3 people died young in the siblings of the patient. Gene sequencing is planned to investigate whether the patient's morbidity is genetic.

Possibility of non-ovarian originated tumor

The gynecological tumor recurrence of the patient was not found in the year after four postoperative chemotherapies since 2012, based on which, the tumor was considered sensitive to chemotherapy. However, 11 months after surgery, we found the protruding-type adenocarcinoma in right hemicolon, and performed operative treatment. According to the pathological findings, the two tumors were homogenous. Therefore, we could take it into consideration that whether it was caused by metastatic carcinoma rather than ovarian tumors. But the patient suffered from gynecological tumor ahead of colon tumors which was found subsequently. However, no examinations like gastroscopy and enteroscopy were performed in the therapeutic process of gynecologic tumor, and the chief complaint in the treatment with general surgery in Zhongshan Hospital was 'stool characters have changed for several years'. So we can consider that the patient's morbidity is not ovarian originated tumor or primary tumor of the reproductive system, and it may be associated with the dissemination and implantation of digestive tract tumor via enterocoelia.

Analysis of two adjuvant therapies

After the surgery in 2012, the patient was treated with chemotherapy four times, in which carboplatin combined with bleomycin was applied. The follow up was done on schedule, during

which, no signs of tumor recurrence was observed at all. After radical operation for carcinoma of colon in Zhongshan hospital, the patient was suggested to use oxaliplatin in combination with 5-fluorouracil as an auxiliary treatment. However the patient refused because of her own reasons. Oxaliplatin is the third generation of platinum anticancer drugs, having strong anticancer activity and wide anticancer spectrum [10]. Targeting DNA, oxaliplatin combines with DNA chains via covalent bonds, and forms inter-strand crosslinks, which results in the blocking of DNA replication and transcription. For advanced colorectal cancer, the effective rate of single drug oxaliplatin in first line treatment was about 20~50%, while in the second-line treatment, the effective rate was even lower (10%). Combined with 5 fluorouracil (5-Fu) or calcium folinate in second line treatment, the effective rate of oxaliplatin reached about 32~58%. However, there is no correlation between postoperative chemotherapy regimens of gynecological surgery and that of general surgery which was planned, thus preliminarily excluding the correlation between two tumors. But for the isogeny of the gynecological tumor and colon tumor, the comparison of their pathological sections remains to be performed.

Therapy and prognosis

Lesion resection is a preferred treatment. It had been 7 years since the total hysterectomy, so double adnexectomy, greater omentum resection, and pelvic lymph node dissection was performed. Subsequently, combined chemotherapy using carboplatin and bleomycin as auxiliary treatment was carried out. So far, no gynecology tumor recurrence and metastasis have been observed in the follow up. But, as the tumor is rare, and there are no similar cases at home and abroad, long-term biological behavior remains to be further confirmed by the accumulation of the cases, and the association between tumor incidence and colon cancer also needs to be clarified.

Acknowledgements

This study was supported by no funding.

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Yu Gu, Obstetrics and Gynecology Hospital Affiliated to Fudan University, 128 Shenyang Road, Yangpu District, Shanghai 200090, China. Tel: +86-15900860784; Fax: +86-21-60513996; E-mail: youngy_1@163.com

References

[1] Chen C, Chow S, Chen RJ, Chen YP, Huang L, Wang L, Lin YH, Huang SC. Primary ovarian cancer at National Taiwan University Hospital: 1980-1989. Int Surg 1993; 79: 48-51.

[2] Fan L. Ovary clinical and pathology. Tianjin: Tianjin Science and Technology Press; 1993. pp. 1998-02.

[3] Naniwadekar MR, Patil N. Extraovarian granulosa cell tumor of mesentery: a case report. Pathol Res Int 2010; 2010: 292606.

[4] Xiaonei D, Yi G, Xie B, Pan F, Huang Y. Clinicopathologic analysis on malignant Wolffian adnexal tumor. Acta Academiae Medicinae Wannan 2010; 29: 422-5.

[5] Lv Y, Wu N, Yu B, Xia C, Zhang R, Wang J, et al. Uterine sex cord-like tumors: a clinical and pathological analysis of 3 cases. Chinese Journal of Clinical and Experimental Pathology 2014; 30: 494-7.

[6] Clement P, Scully R. Uterine tumors resembling ovarian sex-cord tumors. A clinicopathologic analysis of fourteen cases. Am J Clin Pathol 1976; 66: 512-25.

[7] Clement PB, Scully RE. Uterine tumors with mixed epithelial and mesenchymal elements. Semin Diagn Pathol 1988; 5: 199-222.

[8] Clement PB, Young RH, Scully RE. Ovarian granulosa cell proliferations of pregnancy: a report of nine cases. Hum Pathol 1988; 19: 657-62.

[9] Oztekin O, Soylu F, Yigit S, Sarica E. Uterine tumor resembling ovarian sex cord tumors in a patient using tamoxifen: report of a case and review of literature. Int J Gynecol Cancer 2006; 16: 1694-7.

[10] Ye G. Side effects of the combination of Tegafur Gimeracil Oteracil Potassium and Oxaliplatin in the chemotherapy of advanced colon cancer. Strait Pharmaceutical Journal 2013; 25: 281-2.