Repeated recurrence of ovarian sertoli tumor in the laparoscopic port sites with unsuccessful salvage therapies; a case report and review of literature

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Abstract

Objective: Evaluating the role of different salvage therapies in port sites metastasis management in sertoli-leydig cell tumors.

Case presentation: We present a young patient with three times of relapse, on whom, laparoscopic unilateral salpingo-oophorectomy had been performed due to a right persistent complex cyst with 4 cm in diameter. Histopathological findings were in favor of poorly differentiated sertoli-leydig cell tumor and all the recurrences happened at the port site every time. Surgery, chemotherapy and pelvic radiation were applied on her. However, she died during the fourth recurrence of poorly differentiated sertoli-leydig cell tumor about 40 months after initial diagnosis.

Conclusion: Minimally invasive procedures for management of ovarian mass should be done by a specialist person in order to precisely refer for adjuvant chemotherapy, when it is necessary. Pelvic irradiation had almost never been used for sertoli tumors recurrence but for our patient, this was the only way left. This option was not successful either.

Keywords: Sertoli-Leydig tumor, Recurrence, Metastasis, Sex-cord tumor

Introduction

Sertoli-leydig cell tumor (SLCT) of the ovary is attributed to some heterogeneous tumors which includes less than 7% of ovarian malignancies [1] These tumors are often develop in the second and third decades of life, like germ cells tumors but no germ cell component is found in these sex cord-stromal tumors [2]. The most clinical manifestations are unilateral mass with abdominal pain. Nevertheless, virilization caused by androgenic secretion may be seen in some patients [3]. Sertoli tumors are divided into three categories histopathologically as follows: well, moderate and poorly differentiated. On presentation they are usually in stage 1 and the most commonly used treatment is unilateral salpingo-oophorectomy nearly in all cases [4]. Poorly differentiated or more advanced stages are very rare and this rarity leads to challenges in adjuvant therapies in such conditions. Although, based on NCCN Guidelines, adjuvant chemotherapy is recommended [5]. Of course, the results of chemotherapy are not yet known in large volume studies. There are very few reports on the recurrence of SLCT, and a unique consensus to manage it, has not been stated so far. We still do not know how to deal with this particular type of tumor, which ultimately benefits patients. The types of chemotherapy regimen and radiotherapy are unknown. There are no major research studies that are available, even the case reports are very insignificant. Herein, we present a woman with three times relapses in the laparoscopic port sites who did not response to various chemotherapy regimen and pelvic irradiation.

Case report

25-year-old woman was referred to us at joint committee in Gynecology Oncology Department of Tehran University of Medical Sciences, Tehran, Iran, with a large mid-pelvic mass. Information on her past surgical history was revealed during laparoscopic right salpingo-oophorectomy, due to abdominal pain and persistent complex cyst with about 4 cm in diameter on imaging at one of the non-teaching Hospital six months ago. Histopathological diagnosis was poorly differentiated SLCT of the ovary. Unfortunately,
Despite these diagnoses, no adjuvant treatment and surveillance was considered. On her detailed physical examination, a large mobile palpable mass was found just in the umbilicus area which seemed to be adhered to the umbilicus. She didn’t have any symptoms in favor of DE feminization or masculinization as her particular tumor entity. Laboratory hormonal tests were in normal range with only high serum level of CA 125 about 551 U/ml. Our joint committee’s standpoint according to the negative distant metastases on imaging studies, was surgery followed by chemotherapy. So we performed omentectomy, para-aortic and pelvic lymphadenectomy via amidline incision and excised a small part of the fascia beneath the umbilicus in addition to removal the large mass 15&20 cm in size adhering to abdominal wall, penetrating into the abdominopelvic cavity. The remained left ovary and the uterus seemed to be normal. Therefore, were preserved. Aforementioned above, the right ovary had been removed in previous surgery. The microscopic examination revealed a poorly differentiated sertoli tumor with extensive necrosis and heterologous elements (Rabdomyoblast), Figures 1,2 and on immunohistochemistry (IHC) staining the specimen was positive for Inhibin, Calretinin, Vimentin and Myogenin. Whereas it was negative for EMA, CD99 and LCA, also CK was positive in sertoli cells focally. According to histopathologic and IHC findings, recurrence of sertoli tumor was endorsed. Four cycles of BEP chemotherapy regimen containing Bleomycin, Etoposide and Cisplatin were considered based on the recurrence and poorly differentiated grading of the tumor and existence of heterologous elements. She went under close monitoring with exact physical examination and pelvic sonography every three months after completing chemotherapy. After about one year, second recurrence developed. She was undertaken the second debulking surgery due to a large abdominopelvic mass just in the same place of the previous tumor with 8 & 10 cm size. We also performed hysterectomy, appendectomy and left salpingo-oophorectomy at the time.

In fact, debulking surgery was performed during the second relapse. Second relapse was confirmed after pathological examination and six cycles of paclitaxel plus carboplatin were prescribed. During her surveillance, four months after completion chemotherapy with paclitaxel plus carboplatin, three masses were discovered on Magnetic Resonance Imaging (MRI) with sizes of 21mm, 12mm and 23 mm in the left lower quadrant abdominal wall, umbilicus, and cul-de-sac pouch, respectively. After pathologic confirmation of third relapse via sonographic guided fine needle biopsy of abdominal wall mass, irradiation was performed for her, as a salvage therapy. Five months after pelvic irradiation, her abdomen became distant due to severe ascites; multiple tumors inside the abdominopelvic cavity and intestinal obstruction discovered in the spiral scan imaging. She was transferred to the intensive care units to get stabilized for immediate surgery but eventually she died before operation and the leading cause of death was severe resistant acidosis. Pelvic irradiation almost never been used for recurrence of sertoli tumors but for our patient, this was the only option in joint committee consensus due to difficult surgical technique and extensive bowel adhesion which was found in the third surgery. She underwent External Beam Pelvic Irradiation for sequentially periods but this option was not successful either.

Discussion

SCLTs belong to the sex-cord stromal tumors group which almost develop in second and third decades of life, have a good prognosis and low rate of recurrence or distant metastasis. They are generally presented at early stages and fertility sparing surgery is a safe option for these patients. Although the extra ovarian spreading and relapses are rare but exist and may occur during two to three years after initial diagnosis [6,7]. Our case was in her second decade and suffered from poorly differentiated SCLT diagnosed via laparoscopic right unilateral salpingo-oophorectomy. Substantial prognostic factors for recurrence are higher stages and poorly differentiation, as well as heterogeneous elements like muscular, skeletal and endodermal elements [8]. Now a days, laparoscopic surgical methods are used widely for
ovarian mass particularly in young patient and several studies have stated its safety. For example, some of them reported no significant differences in overall survival and disease-free survival between laparotomy and minimally invasive surgical procedure, while others don’t approve such of assertions [9,10]. In addition to the possibility of tumor rupture and its spread into the abdomen and pelvic cavity, port site metastasis is another concern about minimally invasive procedure in malignant ovarian neoplasms [11]. Three relapses occurred in the umbilical port site in our patient, and one of them occurred along with the lateral port site metastasis. In addition to all these explanations, the tumor had never been torn during laparoscopy because it was only 4 cm in size and was removed through a small incision around the umbilicus which was actually created by enlarging the port site hole. On our detailed history, end loop was not used to remove the tumor that may have been the leading cause of repetitive recurrences. Another point which was not much considered was the lack of chemotheraphy following the first surgery despite the poor differentiation of the tumoral cells. Although, the value of adjuvant chemotherapy has not been determined substantially, but it is considered in advanced stages and poor differentiated cells as well as existence of heterologous elements. Our case was in stage one and there were not any heterogeneous elements in histopathological examination but the tumor was high grade and she should have been treated with chemotherapy after surgery. Recurrence often occurs within the two or three years after initial diagnosis and our case experienced recurrence disease only one year after surgery. In recurrence cases, surgery is recommended in the first recurrence. In the first relapse, we performed surgery but one ovary due to its normal intraoperative appearance was preserved and we removed the mass. Only unilateral salpingo-oophorectomy is the best treatment in early stages, which was performed laparoscopically in our patient before advanced surgery is recommended for higher stages, for example bilateral salpingo-oophorectomy plus hysterectomy and omentectomy in case of involvement with pelvic tumors. Dependence of these tumor growth on the sexual hormones, and omentectomy in case of involvement with tumoral tissues. Failure of these tumor growth on the sexual hormones, has not been established but radical surgery and removal of any retained ovarian tissue may be the preferable option in recurrence cases even in younger patients with desire to preserve fertility, because such tumors can be very aggressive. We thought chemotherapy with BEP could be curative after second surgery so did not perform contra-lateral oophorectomy with the aim of maintaining fertility in first recurrence. Carboplatin and Paclitaxel are used successfully as salvage therapy after failure of SLCT. We tried it since all of the other options didn’t work. In addition, based on the unpredictable behavior of these tumors, Fertility sparing surgery is favorable option only in first surgery and should be avoided during the first relapse and complete surgery recommend irrespective of the patient’s age and her desire for fertility in recurrence cases.

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Conflict of interest

All authors have not any conflict of interest to declare.

References


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