OPEN ACCESS
Scholars International Journal of Obstetrics and Gynecology
Abbreviated Key Title: Sch Int J Obstet Gynec
ISSN 2616-8235 (Print) |ISSN 2617-3492 (Online)
Scholars Middle East Publishers, Dubai, United Arab Emirates
Journal homepage: https://saudijournals.com/journal/sijog/home

Case Report

Ovarian Carcinosarcoma: Diagnosis and Treatment –About a Rare Case

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DOI: 10.36348/sijog.2019.v02i08.001 | **Received:** 15.07.2019 | **Accepted:** 22.07.2019 | **Published:** 18.08.2019

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Abstract

Ovarian carcinosarcoma, also called a mesodermal mixed tumor or mixed Mullerian tumor, is a rare ovarian tumor that accounts for less than 2% of ovarian cancers. It is an aggressive tumor that combines a carcinomatous component with a sarcomatous component. Less than 400 cases have been reported in the literature. We report 01 case observed over five years at the department of obstetric and gynecology of the university hospital Hassan II. Ovarian carcinosarcoma has a worse prognosis than ovarian epithelial tumors. Its rarity explains that there is no consensus on its management. There is very little data available. The only prognostic factor found is the initial stage. The surgical management is a determining factor for the survival of the patients, this one must be as complete as possible. The sensitivity to the chemotherapy is lower, is about 20%. The survival at five years is lowered when the compared to epithelial tumors of the ovary.

Keywords: Ovarian carcinosarcoma, prognostic factors, management.

INTRODUCTION

Malignant-mixed mullerian tumors (MMMT)/carcinosarcomas are rare tumors and pathologically consist of malignant epithelial and malignant mesenchymal components. Of these cases, ovarian carcinosarcomas are very rare and account for only 1%–2% of all malignant ovarian tumors. With rare exceptions, attempts to run prospective clinical trials in this disease have been elusive, and randomized clinical trials will probably never be performed. Therefore,the optimal treatment of OCS remains uncertain due to this tumor's rare occurrence. Many cases of OCS undergo surgical treatment and chemotherapy, similar to epithelial ovarian cancer.

We report 01 case observed over five years at our departement of gynecology in the university hospital of FES.

CASE PRESENTATION

A 68-year-old woman presented with an externally huge tumor in the lower abdomen. The tumor was restricted and reached 5 cm above the navel. Imaging findings, including computed tomography and magnetic resonance imaging, revealed a multilocular cyst tumor with a diameter of 18 cm, imaging also showed that the patient had massive ascites. The preoperative serum level of cancer antigen 125 (CA125) was elevated to 437.3 U/ml (normal range: < 35.0), whereas the carcinoembryonic antigen (CEA), cancer antigen 19–9 (CA19–9) and squamous cell carcinoma (SCC) values were within the respective normal ranges.

Total abdominal hysterectomy, bilateral adnexectomy, omentectomy,and biopsy of parieto-colonic gutter was performed. The lymphadenectomy (pelvic and para-aorta), was not realized because of the android obesity. At surgery, massive hemorrhagic ascites of 3000 ml was present in the perinatal cavity. The ruptured tumor arising from the right ovary was found firmly adhered to the sigmoid colon, transverse colon, and mesentery of small intestine.

The postoperative course was uneventful. Histologically, most of the tumor showed undifferentiated pleomorphic sarcoma, in which tumor cells of various forms with strong nuclear atypia grow complicatedly. The immunohistochemical analysis showed that the sarcomatous component was positive for vimentin, alpha SMA and CD10; the Ki-67 (MIB-1) index was 60%. The carcinomatous component

comprised squamous cell carcinoma. Its immunohistochemical analysis showed positivity for AE1/AE3.The final diagnosis was OCS classified as at least (pT1c) according to the International Federation of Gynecology and Obstetrics (FIGO) 2014 classifications with squamous cell carcinoma as the carcinomatous component.

DISCUSSION

It has been suggested from previous studies that ovarian carcinosarcoma is a relatively rare clinicopathologic entity that has a tendency to affect an elderly population and is associated with poor prognosis. In the current series, we found that carcinosarcomas represented 4% of the total number of epithelial ovarian carcinomas, which to our knowledge is a higher proportion than previously described [1]. Carcinosarcomas of the female genital tract are often found after menopause at a median age of 60 to 70 years old. More than two-thirds of patients with OCS are diagnosed at an advanced stage [2].

Depending on the sarcomatous component, we define two types: either the sarcomatous component is normally present in the ovary we will speak about CSO counterpart, or the component is formed of elements usually absent (cartilaginous tissue, bone, muscle fibers striated ...) we will talk about heterologous CSO. The heterologous type is most often described [3]. The composition during the course of the disease varies: at the time of diagnosis, the carcinomatous elements are in the majority, whereas in case of recidivism, the sarcomatous elements predominate [4, 5]. OCS has a worse survival rate than high-grade ovarian cancer at the same FIGO stage, with a median overall survival ranging from 7 to 27 months [6].

The clinical presentation of CSO is nonspecific. The most common symptom is abdominal distension [7]. It may be associated with abdominal pain,transit disorders and impairment of general condition [5]. Very often, the diagnosis is made at an advanced stage of the disease [4], [5]. Metastatic localization nor do they differ from those of epithelial tumors the ovary [5].

Rustin and Brown have studied the interest of the dosage of CA 125 in the CSO [8], [9]. It is increased in 75 to 85% of case [5]. Although not validated, it seems to be a marker interesting in the therapeutic evaluation, in the absence clinical or radiological criteria.

The optimal treatment of OCS remains uncertain due to this tumor's rare occurrence. Many cases of OCS undergo surgical treatment and chemotherapy, similar to epithelial ovarian cancer [10]. In one of the larger studies, including 50 patients with OCS, the disease-free survival for patients with complete resection was 19 months. In contrast, the disease-free survival of patients with optimal surgery (< 1 cm residual disease) was 10 months, while that with suboptimal surgery (≥1 cm residual disease) was 5 months. The overall survival of complete resection and optimal and suboptimal surgery is reportedly 47, 18 and 8 months, respectively [11]. Optimal surgical cytoreduction, including total abdominal hysterectomy, bilateral adnexectomy, omentectomy, pelvic and para-aortic lymph node dissection, and tumor debulking is important for improving the prognosis of OCS. Following debulking surgery for OCS, adjuvant chemotherapy is typically recommended. However, there is no consensus regarding the most effective regimen for such a rare malignancy [12].

Multiple chemotherapeutic regimens have been evaluated with modest response rates ranging from 12– 100%. The trials are difficult to evaluate due to small numbers of patients, multiple treatment regimens, and occasional use of radiation therapy. Some of the chemotherapy regimens evaluated in the past include: PAD (cisplatin, doxorubicin, dacarbazine); VAC (vincristine, dactinomycin, and cyclophosphamide); CAP (cyclophosamide, doxorubicin, cisplatin); doxorubicin/ifosfamide, cisplatin/ifosfamide, and carboplatin/taxol. In OCS, similar to epithelial ovarian cancer, platinum-based chemotherapy is considered a key drug. Combination chemotherapy, such as carboplatin and paclitaxel or ifosfamide, exhibits a higher response rate than single-agent platinum chemotherapy [13]. Several studies have described the clinical effectiveness of combination chemotherapy with carboplatin and paclitaxel in ovarian squamous cell carcinoma [14]. Given the above findings, combination chemotherapy with carboplatin and paclitaxel was selected in the present case. Ultimately, the present patient died after her 2nd chemotherapy session following a pulmonary embolism.

CONCLUSION

The CSO is a special, rare entity with a poor prognosis. Very few cases have been reported in the literature. Two histological types are described: the heterologous type and the homologous type, but without affecting the prognosis. Indeed, the only prognostic factor found in the different studies is the initial stage. Five-year survival is lowered when compared to TEO. The slightest sensitivity to chemotherapy offers surgery a key role, this one must be as complete as possible.

REFERENCES

1. Bicher, A., Levenback, C., Silva, E. G., Burke, T. W., Morris, M., & Gershenson, D. M. (1995). Ovarian malignant mixed müllerian tumors treated with platinum-based chemotherapy. *Obstetrics & Gynecology*, *85*(5), 735-739.
2. del Carmen, M. G., Birrer, M., & Schorge, J. O. (2012). Carcinosarcoma of the ovary: a review of the literature. *Gynecologic oncology*, *125*(1), 271-277.

3. Hellström, A. C., Tegerstedt, G., Silfverswärd, C., & Pettersson, F. (1999). Malignant mixed müllerian tumors of the ovary: histopathologic and clinical review of 36 cases. *International Journal of Gynecological Cancer*, *9*(4), 312-316.
4. Ariyoshi, K., Kawauchi, S., Kaku, T., Nakano, H., & Tsuneyoshi, M. (2000). Prognostic factors in ovarian carcinosarcoma: a clinicopathological and immunohistochemical analysis of 23 cases. *Histopathology*, *37*(5), 427-436.
5. Amant, F., Vloeberghs, V., Woestenborghs, H., Moerman, P., & Vergote, I. (2003). Transition of epithelial toward mesenchymal differentiation during ovarian carcinosarcoma tumorigenesis. *Gynecologic oncology*, *90*(2), 372-377.
6. Berton-Rigaud, D., Devouassoux-Shisheboran, M., Ledermann, J. A., Leitao, M. M., Powell, M. A., Poveda, A., ... & Kim, J. W. (2014). Gynecologic Cancer InterGroup (GCIG) consensus review for uterine and ovarian carcinosarcoma. *International Journal of Gynecologic Cancer*, *24*(Supp 3), S55-S60.
7. Eichhorn, J. H., Young, R. H., Clement, P. B., & Scully, R. E. (2002). Mesodermal (müllerian) adenosarcoma of the ovary: a clinicopathologic analysis of 40 cases and a review of the literature. *The American journal of surgical pathology*, *26*(10), 1243-1258.
8. Rustin, G. J., Nelstrop, A. E., Bentzen, S. M., Bond, S. J., & McClean, P. (2000). Selection of active drugs for ovarian cancer based on CA-125 and standard response rates in phase II trials. *Journal of clinical oncology*, *18*(8), 1733-1739.
9. Rustin, G. J., Nelstrop, A. E., McClean, P., Brady, M. F., McGuire, W. P., Hoskins, W. J., ... & Lambert, H. E. (1996). Defining response of ovarian carcinoma to initial chemotherapy according to serum CA 125. *Journal of Clinical Oncology*, *14*(5), 1545-1551.
10. Rutledge, T. L., Gold, M. A., McMeekin, D. S., Huh, W. K., Powell, M. A., Lewin, S. N., ... & Mannel, R. S. (2006). Carcinosarcoma of the ovary—a case series. *Gynecologic oncology*, *100*(1), 128-132.
11. Rauh-Hain, J. A., Growdon, W. B., Rodriguez, N., Goodman, A. K., Boruta II, D. M., Schorge, J. O., ... & del Carmen, M. G. (2011). Carcinosarcoma of the ovary: a case–control study. *Gynecologic oncology*, *121*(3), 477-481.
12. Shylasree, T. S., Bryant, A., & Athavale, R. (2013). Chemotherapy and/or radiotherapy in combination with surgery for ovarian carcinosarcoma. *Cochrane Database of Systematic Reviews*, (2).
13. Duska, L. R., Garrett, A., Eltabbakh, G. H., Oliva, E., Penson, R., & Fuller, A. F. (2002). Paclitaxel and platinum chemotherapy for malignant mixed mullerian tumors of the ovary. *Gynecologic oncology*, *85*(3), 459-463.
14. Eltabbakh, G. H., Hempling, R. E., Fernando, O., & O'Neill, C. P. (1998). Remarkable response of primary squamous cell carcinoma of the ovary to paclitaxel and cisplatin. *Obstetrics & Gynecology*, *91*(5), 844-846.