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Debulking surgery for advanced stage ruptured ovarian carcinosarcoma with secondary hemoperitoneum and rectosigmoidian invasion. A case report and literature review

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Abstract

Ovarian carcinosarcomas are rare gynecological malignancies with high biological aggressiveness and poor outcomes when compared to their epithelial counterparts. However, it seems that the only way to offer a chance for cure for these patients is an aggressive surgical approach in order to achieve complete cytoreduction to no visible residual disease. We present a 57 year old woman who presented for diffuse pelvic pain and constipation and who was diagnosed with a large ruptured pelvic mass invading the recto-sigmoid colon. The patient was submitted to surgery, a total hysterectomy with bilateral adnexectomy en bloc with recto-sigmodian resection, pelvic and para-aortic lymph node dissection being performed. The histopathological studies confirmed the presence of an ovarian carcinosarcoma.
Keywords: *debulking, ovarian carcinosarcoma, rectosigmoidian invasion*

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Introduction

Ovarian carcinosarcomas are also known under the name of mixed mullerian tumors and represent up to 4% of all gynecologic malignancies[1,2]. Their characteristic consists in the association between malignant stromal and epithelial elements and they are best considered as metaplastic epithelial carcinosarcomas[3]. The particularity of these tumors is the biological aggressiveness of the cells, most patients being diagnosed in advanced stages of the disease when disseminated lesions are already present. In consequence, optimally cytoreductive surgery is hard to be achieved, most patients being submitted to suboptimal resections, with poor long term results[2]. Moreover, it has been showed that up to 90% of cases presented disseminated lesions at the time of initial diagnosis, including the cases in International Federation of Gynecology and Obstetrics (FIGO) stages III or IV. This fact is translated in very poor long term prognosis, the rate of 5-year overall survival ranging between 7-20%[4-6]. The particular poor outcome in association with the rarity of cases is responsible for the fact that most studies conducted on cases with ovarian tumors consider this histopathological type as exclusion criteria.

Case report

A 57 year old patient presented for diffuse pelvic pain and constipation. The clinical examination revealed the presence of peritoneal irritation signs. The vaginal examination revealed the presence of a large pelvic mass palpable in the both Douglas pouches while the rectal examination revealed the presence of narrowed rectal ampula. The patient was submitted to a computer tomography scan which revealed a large pelvic mass invading the recto-sigmoidian colon associated with free peritoneal fluid. The patient was submitted to emergency exploratory laparotomy. Intraoperatively, a large ruptured pelvic mass invading the recto-sigmoidian colon, with disseminated peritoneal lesions and secondary hemoperitoneum was found. A total hysterectomy en bloc with bilateral adnexectomy en bloc with recto-sigmoidian resection, pelvic and pa-

ra-aortic lymph node dissection, parietal and pelvic peritonectomy was performed. Cytoreduction to no residual disease was achieved (Figures 1-4).

The distal sigmoidian loop was exteriorized in left colostomy. The patient was discharged in the 5th post-operative day. The histopathological studies revealed the presence of an ovarian carcinosarcoma. One month after surgery the patient was submitted to platinum based adjuvant chemotherapy.

Currently the patient has completed the sixth cycle of adjuvant chemotherapy regimen and reports a good quality of life. No signs of recurrent disease were found at six months of follow up.

Discussion

Carcinosarcomas of the ovary are poor prognostic tumors, with low rates of long term survival. Patients diagnosed with ovarian carcinosarcomas present similar symptoms to those diagnosed with epithelial ovarian cancer. However, the prognostic is significantly poorer, with low rates of 5 year overall survival especially if advanced stage of disease is present at the time of diagnosis. Due to the fact that these cases are very rare, no standard therapeutic protocol has been established until present. Therefore, the existence of similar points between ovarian carcinosarcomas and ovarian adenocarcinomas enabled the surgeons worldwide to propose debulking surgery as the only potential curative therapeutic option. The most important prognostic factors in these cases remain the stage at initial diagnosis, the ability to perform an optimal cytoreduction and the type of adjuvant chemotherapy[2].

In order to compare the long term outcomes of patients with ovarian carcinosarocma with those reported in cases diagnosed with epithelial ovarian adenocarcinomas in similar stages, Rauh-Hain et al. conducted a study on 50 patients diagnosed with ovarian carcinosarcoma. The reported results were compared to the ones reported by 100 patients diagnosed with epithelial ovarian tumors in similar stages, each case being compared to two control cases. Optimal cytoreduction rates were similar between cases and control cases (i.e. 74% versus 81%, p=0.6). When it comes to the platinum, the chemotherapy response is achieved. The overall response rate was 62% among patients diagnosed with carcinosarcomas, significantly lower compared to the one reported by the epithelial adenocarcinoma cases (83%, p=0.03). In the meantime, platinum sensitivity at six months from completion of adjuvant chemotherapy reported a significant decrease among cases with carcinosarcomas. As for the time to recurrence and median overall survival rates, both parameters reported significantly lower values for patients diagnosed with carcinosarcomas (i.e. the median disease free survival was 11 months versus 16 months, p=0.002 while the median overall survival was 24 months versus 41 months, p=0.002). In multivariate analysis sarcomatous subtype and suboptimal cytoreduction were significant predictive factors for poor outcome. Among patients with carcinosarcomas,

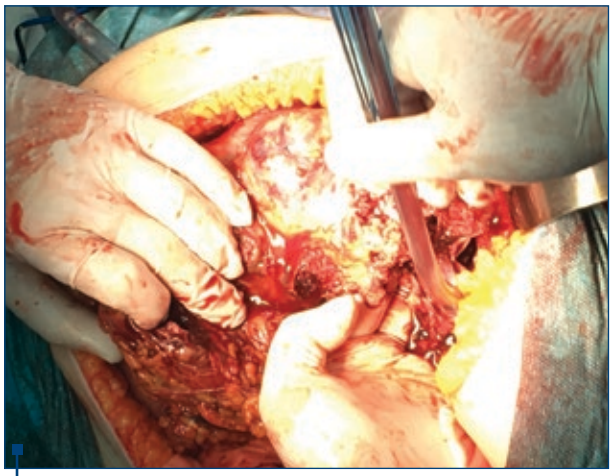

Figure 1. Large ruptured ovarian tumor with secondary hemoperitoneum

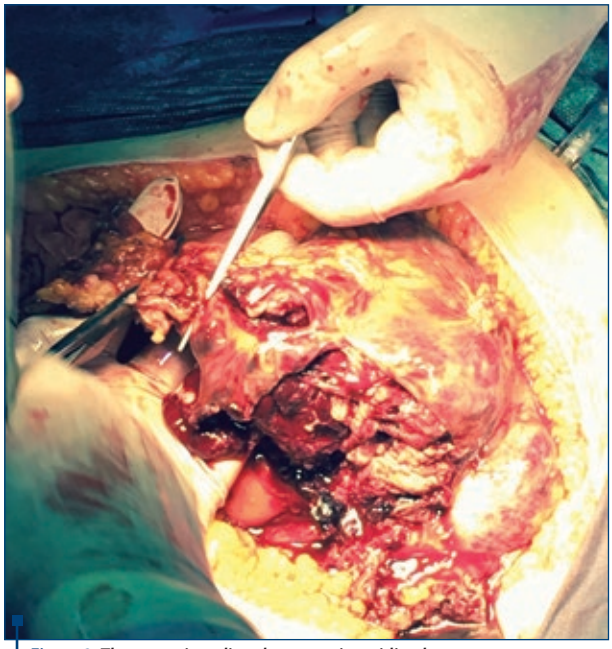

Figure 2. The tumor invading the recto-sigmoidian loop

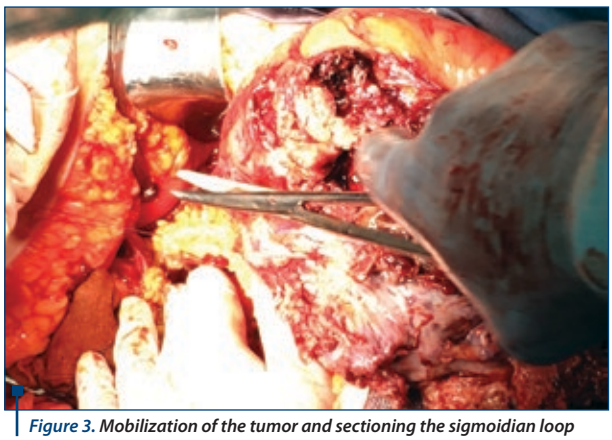

Figure 3. Mobilization of the tumor and sectioning the sigmoidian loop

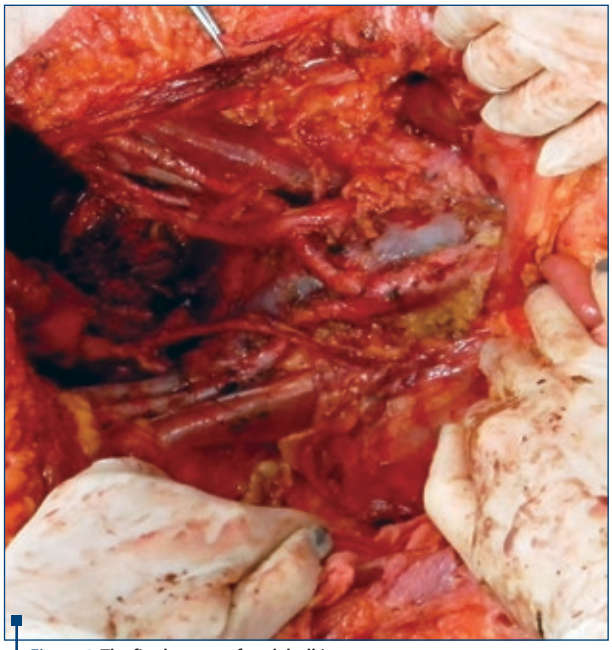

Figure 4. The final aspect after debulking surgery

the overall prognostic was significantly influenced by the completeness of cytoreduction[7].

In another study conducted by the same team regarding the prognostic factors in cases diagnosed with ovarian, fallopian tube and peritoneal carcinosarcomas, the main prognostic factor predicting long term survival remained completeness of cytoreductive surgery, followed by the stage and age at diagnosis. Other incriminated factors were related to the histopathological characteristics of the tumor, a sarcomatous component higher than 25% being associated with a poor prognosis[8]. In the meantime, molecular studies suggest that p53 overexpression, as well as increased expression of vascular endothelial growth factor and Ki67 expression[9-11].

Another case-control study which compared the outcomes of the patients diagnosed with ovarian sarcomas to the ones reported by the patients diagnosed with similar stage epithelial ovarian adenocarcinomas was the one conducted by Bacalbasa and contributors and published in 2014. The authors identified 11 patients diagnosed with ovarian carcinosarcomas in "Dan Setlacec" Center of General Surgery and Liver Transplantation, Fundeni Clinical Institute between 2002 and 2013. Among these cases, carcinosarcomas were reported in nine cases, optimal debulking surgery being achieved in four cases. When the long term outcomes of the patients were compared to the ones reported by similar cases diagnosed in similar stages but with epithelial ovarian adenocarcinomas, a worse prognostic was reported among all FIGO stages[12].

One of the most representative studies which focused on the subject of the effectiveness of cytoreductive surgery and the impact of adjuvant platinum based chemotherapy on the overall survival comes from Amelia Jernigan et al. and included 47 patients diagnosed with ovarian carcinosarcomas. The median age at diagnosis was 47 years while the classification according to FIGO stage identified 11 patients with stage I-II tumors, 27 patients with stage III tumors and 6 cases with stage IV tumors, while in the remnant 3 cases data regarding preoperative status were not available. Cytoreduction to no residual disease was achieved in 51.1% of cases while other 19.1% of patients were debulked to less than 1 cm residual tumor and the remnant 29.8% were suboptimally debulked (i.e. a remnant tumoral volume larger than 1 cm). Lymph node dissection was performed in 27.7% of cases. When it comes to the adjuvant therapy, it consisted in platinum based chemotherapy in 46.8%, non-platinum based chemotherapy in 27.7%, radiation therapy in 6.4% and unknown regimen in 19.1% of cases. In univariate analysis, overall survival was significantly influenced by patients' age, stage at diagnosis and completeness of cytoreduction. In multivariate analysis, age, stage at diagnosis and cytoreductive status significantly impacted on the overall survival. When the regimens of adjuvant chemotherapy were compared, a slightly improved outcome was reported in cases submitted to platinum based chemotherapy. However the difference was not statistically significant[13].

According to a recent European Society for Medical Oncology guideline, if ovarian carcinosarcomas is suspected, surgery should be planned as for any other suspect ovarian malignancy, the aim of the procedure being to achieve no residual disease. The treatment should be continued with adjuvant chemotherapy in all cases, even if FIGO stage I lesions are diagnosed. However, the treatment dilemma refers to the type of adjuvant chemotherapy: whether these tumors should be treated as epithelial tumors (i.e. with platinum and taxanes chemotherapeutic regimens) or whether to include ifosfamide and anthracycline[14].

A significant particularity of our case was also the one that the patients presented with a ruptured lesion, with secondary hemoperitoneum. To the best of our knowledge, only few cases have been reported so far. Another case of gynecological carcinosarcoma rupture presenting with hemoperitoneum comes from Gupta and Jenison and was published in 2012. However, their case referred to a fallopian tube carcinosarcoma in a 57 year old postmenopausal woman who presented for lower abdominal pain. The patient was submitted to surgery, a total hysterectomy with bilateral adnexectomy, omentectomy, pelvic and para-aortic lymph node dissection being performed. The authors reported no residual disease. Postoperatively the patient underwent six cycles of doxorubicin, cisplatinand paclitaxel and was diagnosed with no recurrent disease at eight months after surgery[15].

Conclusions

Ovarian carcinosarcomas are rare gynecologic malignancies with poor outcome due to the biological

aggressiveness of these cells. This aspect is reflected throughout all FIGO stages when comparison to epithelial adenocarcinoma is performed. Although reported cases are rare and no other standard therapeutic protocol has been reported until present, it seems that debulking surgery or no residual disease followed by adjuvant chemotherapy represent the best therapeutic strategy. ■

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