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# OVARIAN CARCINOSARCOMA: ABOUT A CASE AND REVIEW OF LITERATURE

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## ABSTRACT

Ovarian carcinosarcoma is one of the rarest histological subtypes of ovarian cancer. There are approximately 1-4% of all ovarian cancers. Given the rarity of this histological type, management is often extrapolated from the experience of ovarian adenocarcinoma. The prognosis seems to be more dismal than adenocarcinomas, with a median survival of less than 18 months, very variable according to the studies and the stages of the disease. The histological, homologous or heterologous subtype is not associated with a change in prognosis. On the other hand, the stage of the disease at diagnosis, the age of the patient, the complete surgery, seem to affect survival. Response rate to chemotherapy is approximately 20%. We report a case of 73-year-old female patient, with an ovarian carcinosarcoma stage IIIC.

**WORDSKEY:** Ovarian carcinosarcoma; malignant mixed mullerian ovarian tumor; debluking surgery; chemotherapy.

## INTRODUCTION

Ovarian carcinosarcoma (CS), also called mixed mesodermal tumor or mixed Mullerian tumor, is a rare, aggressive ovarian tumor, approximately 1-4% of all ovarian cancers.[1] less than 400 cases have been reported in the literature.[2] It is characterized by the combination of a carcinomatous component and a sarcomatous component. It most often occurs at the level of the body of the uterus. The ovary, cervix and vagina are more rarely affected. We report in our article a case observed and supported at the Souissi maternity hospital and the Moulay Abdellah hospital of oncology CHU Rabat-Sale

### Case presentation

We report the case of a 73-year-old. Hypertensive woman treated with amlodipine, gravida 3 para 3 (G3P3), postmenopausal, who had experienced 2 months of progressive abdominal pain and abdominal distension with pollakiuria and episodes of acute urinary retention, admitted to the Obstetrics and Gynecology Department of the University Hospital of Rabat. She reported having a stable weight, and her vital signs were within normal range She had no relevant past medical history. Physical examination showed a pelvic mass and a right inguinal lymphadenopathy.

Pelvic ultrasound revealed a polylobed and heterogeneous right pelvic mass containing solid parts hypervascular of about 11.5 cm in size (Figure 1). pelvic MRI showed a mixed solidocystic mass seemed to emerge through the right ovary, with malignant aspect (Figure 2). The preoperative serum level of cancer antigen 125 (CA125) was elevated just to 66.40 U/ml. atrophic cervicovaginal smear.

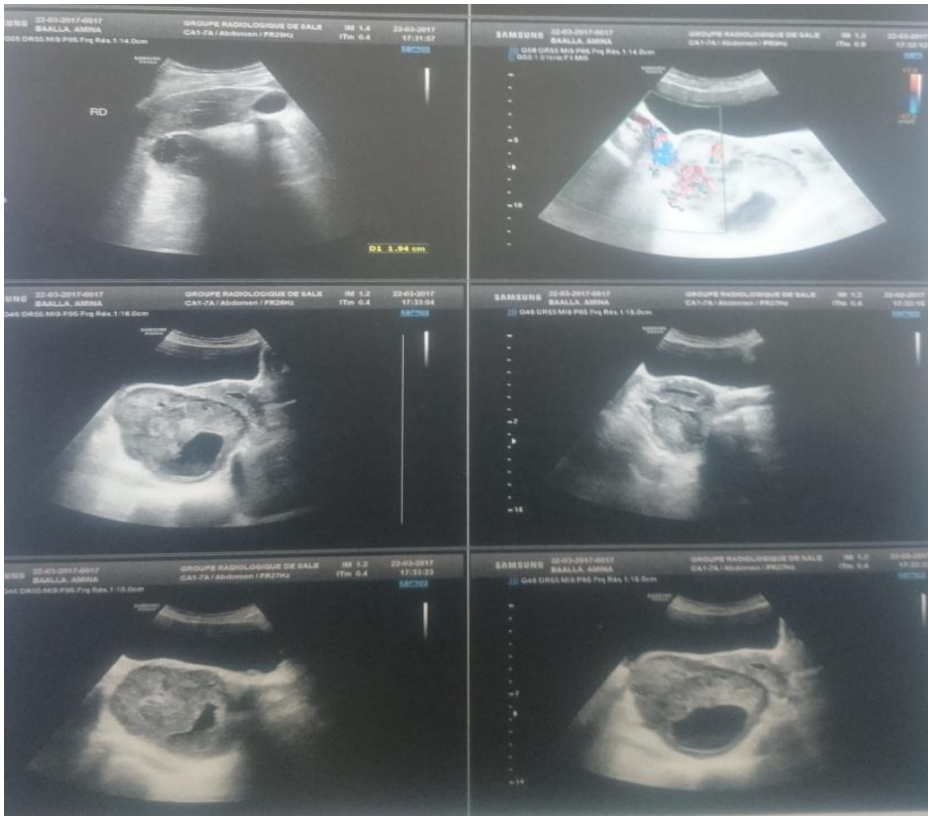

**Figure 1: A pelvic ultrasound shows a right lateral pelvic mass with double cystic and tissue component, this lesion is polylobed measuring 11.5 cm for major axis.**

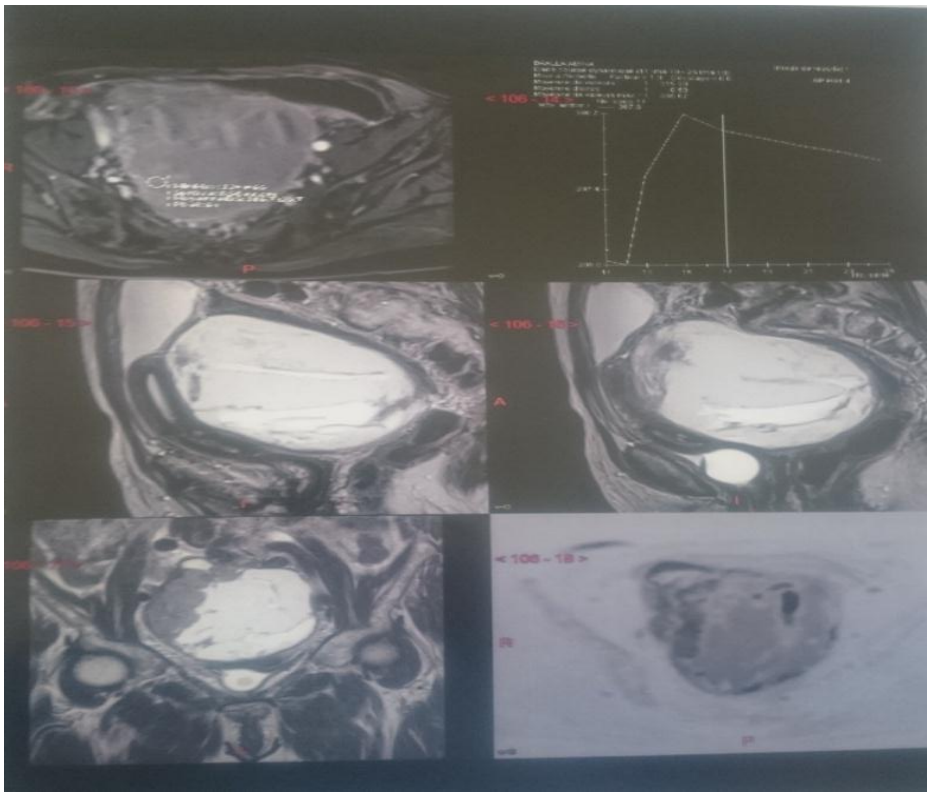

**Figure 2: MRI findings mixed right solidocystic pelvic tumor, which the aspect was malignant.**

Under the suspicion of ovarian malignancy, it was decided to conduct an explorative laparotomy (first look) showing on a 12 / 10cm right ovarian tumor densely adherent to the uterus, bladder, in anterior, and adhered to the sigmoid colon, transverse colon, mesentery of small intestine in posterior with diffuse carcinomatosis, a

cytoreduction of tumor was performed with biopsies of great omentectum and parietal peritoneum, and peritoneal washings for staging.

The histological examination showed heterologous malignant mixed mullerian tumor of the ovary (mixed malignant mullerian tumor) with peritoneal and epiploic invasion, and positive peritoneal washings.

The patient underwent optimal debulking surgery (second look) including sub-total hysterectomy, bilateral salpingo-oophorectomy omentectomy and appendicectomy, para-aortic and pelvic lymphadenectomy. The postoperative course was uneventful.

The final diagnosis was ovarian carcinosarcoma classified as stage IIIC (pTN1M0) according to the International Federation of Gynecology and Obstetrics (FIGO) 2014 classifications. an apparent early stage cancer is upstaged because of nodal disease confirmed. Postoperatively, the patient started treatment with chemotherapy (paclitaxel and carboplatin) as adjuvant therapy but without efficacy. Patient dies 12 months after diagnosis.

## DISCUSSION

Carcinosarcoma, also called mixed mesodermal tumor or mixed malignant mullerian tumor (MMMT), is an anatomopathological entity associating a sarcomatous component with a carcinomatous component. It most often occurs in the body of the uterus, while the ovary, cervix and vagina are more rarely affected.

Currently, several scientific researchers agree that the majority, but not all, of ovarian CS cells have a monoclonal origin, derived from the same cell, and that the carcinomatous component is the primum movens of these cancers. Indeed, from the work carried out on the uterine CS, the same mutation, TP53, was found on the two components of the CS demonstrating their common origin.[3]

Depending on the sarcomatous component, two types are defined: either that the sarcomatous component is normally present within the ovary, we will then speak of homologous CS; or that the component is made up of elements usually absent (cartilage tissue, bone, striated muscle fibers, etc.), we will then speak of heterologous CS. The heterologous type is most often described.[4] The tumor composition during the disease varies: at the time of diagnosis, the carcinomatous elements are predominant, while in the event of recurrence, the sarcomatous elements predominate.[5,6]

Our patient had the heterologous type, the most frequent one.

The clinical presentation frequently resembles that of an ovarian adenocarcinoma. It would appear, however, that the age at diagnosis would be somewhat higher, with a median age at diagnosis of 60-70 years. The CS affects women, most often nulliparous.

Symptoms include abdominal pain, early satiety, functional gastrointestinal disturbances. Ascites is also found, rather in the advanced stages. Hepatic or pulmonary metastases are rarely present, unlike uterine localizations.[1]

The interest of the CA125 dosage in CS has been studied.[7,8,9] It is increased in 75 to 85% of cases.[7,11] Although not validated, it seems to be an interesting marker in therapeutic evaluation in the absence of clinical or radiological criteria.

The discovery of CS in our patient was at a very advanced age with fast clinic evolution, an apparent early stage cancer is upstaged only because of nodal disease confirmed (IIIC)

The scarcity of CS explains why there is no consensus on its management. There is very little data.

Debulking surgery is also the benchmark in ovarian localization, for tumors that can be resected immediately. Exploratory laparoscopy immediately for staging is recommended. There is no prospective series studying the impact of complete surgery on survival, but retrospective studies tend to show a benefit in case of complete surgery.[11] Rauh Hain et al. show the interest of complete surgery compared to suboptimal surgery with a benefit in recurrence-free survival and overall survival.[12,15]

This procedure includes hysterectomy, bilateral salpingooophorectomy, omentectomy, and appendectomy, para-aortic and pelvic lymphadenectomy to the left renal vein. Peritoneal, para-colic, subdiaphragmatic and minor pelvis biopsies are recommended. In individual cases may be given preoperatively three or six cycles of chemotherapy in order to achieve an optimal surgical result. A retrospective study by LuCHetal, supported that cytoreduction to no gross and no more than 1 residual tumors trend toward an improved survival, even though not statistically significant (P=0.144 and 0.137, respectively).[21] The operative approach of these tumors should resemble the management of EOC, and should be under taken in specialized institutions.[22,23] The role of lymphadenectomy in the management of OCS has not yet been clarified due to the rarity of the disease. In a recent study, a cohort of 363 patients with early stage OCS was divided into two groups; 186 patients underwent lymphadenectomy, whereas 177 did not, respectively.[24] Multivariate analysis identified an independent positive association of early American Joint Committee on Cancer (AJCC) T, and lymphadenectomy with OS. A higher survival benefit was observed in the group of the patients who underwent lymphadenectomy

than in those who did not in AJCCT2 (P=0.002 for OS); never the less, but there was no statistically significant difference between either group in AJCCT1 disease (P=0.582 for OS). in the adjuvant situation, chemotherapy is recommended after complete surgery. The data are essentially retrospective, the large prospective studies, in particular those of the GOG (Gynecologic Oncology Group) generally exclude these anatomo-pathological sub-localizations.

A doublet based on platinum salt is used. The data are contradictory as to the doublet of chemotherapy used. Ifosfamide is the drug that has been shown to be effective, and should remain the preferred drug used with cisplatin. However, a retrospective study shows the interest of paclitaxel associated with carboplatin in survival without recurrence,[13] and this is the case of our patient. Doxorubicin has not been shown to be effective. Regardless of the platinum doublet regimen used, relapse-free survival is significantly worse for carcinosarcomas compared to adenocarcinomas.

A recent study shows that the main prognostic factors are the FIGO stage, complete adjuvant chemotherapy, and the predominant epithelial component.[14,15]

Regarding radiotherapy, Data are lacking in this histological type, especially due to the advanced stages, and the peritoneal involvement making radiotherapy inaccessible. In the very early stages with localized disease, the effectiveness of radiotherapy is not known. Radiotherapy must remain a weapon in the event of palliative, analgesic or decompressive treatment.[15]

CS is an aggressive tumor, survival at five years varies from 6 to 30%.[4,5,16,17,18] The initial stage is the only prognostic factor found in the various studies.[5,17,19] The more advanced the stage, the worse the prognosis. Size, histological type (heterologous or homologous), age are not involved in the prognosis.

At the same stage, CS has a worse prognosis than EOT (Epithelial ovarian tumor). A study carried out a comparison between these two tumors, found a significant difference on the median survival (8.2 versus 20.7 months, p <0.0001) and progression-free survival (6.4 versus 12 months, p = 0.001), and five-year survival is in favor of EOT.[18]

## CONCLUSION

The SC 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 impact on prognosis. Indeed, the stage of the disease at diagnosis, the age of the patient, the complete surgery, seem to affect survival.[20]

The slightest sensitivity to chemotherapy offers surgery an essential place, which must be as complete as possible.

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