International Journal of Academic Health and Medical Research (IJAHMR)
ISSN: 2643-9824
Vol. 5 Issue 4, April - 2021, Pages: 72-74

Carcinosarcoma Of The Ovary: A Case Report And Review Of The Literature

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Abstract: *Ovarian carcinosarcoma (OCS) also called mixed mesodermal tumor or mixed mullerian tumor with a double component: epithelial and sarcomatous rare gynecological tumor that represents less than 2% of ovarian cancers. In the genital tract, it is most often found in the uterus. The ovary, cervix or vagina are more rarely affected. Carcinosarcoma affects women between the ages of 60 and 70. The main prognostic factor found is the initial stage, most often advanced: At diagnosis, more than 90% of ovarian carcinosarcomas have a locoregional extension. The surgical management is a determining factor for the survival of patients. Carcinosarcoma is an aggressive tumor, the median survival is 7 to 27 months for ovarian carcinosarcomas. Relapse occurs mainly in the first year. FIGO stage is the most important prognostic factor. Complete surgery, advanced age, grade of the sarcomatous component, and adjuvant chemotherapy are found in the few published retrospective studies. The response rate to chemotherapy is about 20%. Our objective is to report a case of ovarian carcinosarcoma while performing a literature review.*

Keywords :Ovarian carcinosarcoma, initial stage, surgery, prognostic factor, chemotherapy.

RESUME: Le carcinosarcome ovarien appelé également tumeur mixte mésodermique ou tumeur mixte mullérienne à double composante : épithéliale et sarcomateuse rare tumeur gynécologique qui représente moins de 2% des cancers de l'ovaire. Au niveau du tractus génital, elle siège le plus souvent au niveau de l'utérus. L'ovaire, le col ou le vagin sont plus rarement atteints. Le carcinosarcome intéresse les femmes plutôt entre 60 et 70 ans. Le facteur pronostique principal retrouvé est le stade initial, le plus souvent avancé: Au diagnostic, plus de 90% des carcinosarcomes ovariens ont une extension locoregionale . La prise en charge chirurgicale est un facteur déterminant pour la survie des patientes. Le carcinosarcome est une tumeur agressive, la médiane de survie est à 7 à 27 mois pour les carcinosarcomes ovariens. Les rechutes surviennent essentiellement dans la première année. Le stade FIGO est le plus important des facteurs pronostiques. La chirurgie complète, l'âge avancé, le grade de la composante sarcomateuse et la chimiothérapie adjuvante sont retrouvés dans les rares études rétrospectives publiées. Le taux de réponse à la chimiothérapie est de 20% environ. Notre objectif est de rapporter un cas de carcinosarcome ovarien tout en effectuant une revue de littérature.

MOTS-CLEFS: Carcinosarcome ovarien, stade initial, chirurgie, facteur pronostique, chimiothérapie.

INTRODUCTION:

Ovarian carcinosarcoma (OSC), also known as mixed mesodermal tumor or mixed mullerian tumor, is a rare, and aggressive ovarian tumor that accounts for less than 2% of ovarian cancers [1]. Less than 400 cases have been reported in the literature. It is characterized by the association of a carcinomatous component and a sarcomatous component.

We report the case of an ovarian carcinosarcoma observed in the obstetric gynecology department of Hassan II University Hospital in Fes.

clinical case:

Mrs. N.B, 40 years old, operated for a goiter, nulligest, always regulated, who consulted for pelvic pain of the heaviness type evolving since 3 months associated with a progressive increase of the abdominal volume, without associated gynecological or extra-gynecological signs. The whole evolving in a context of apyrexia and conservation of the general state.

Clinically, the patient was in good general condition, OMS = 1, BMI = 21.5.

The gynecological examination was not done, the patient said she was a virgin.

Abdominal examination: supple abdomen, no defenses or contractures, diffuse abdominal sensitivity, with palpation of a large abdominal-pelvic mass, of hard consistency. Presence of a sloping dullness.

The rest of the somatic examination was unremarkable.

Ultrasound and CT scan show bilateral ovarian solid cystic tumor masses measuring 13*11cm on the right and 6*3 cm on the left, in contact with the uterus and the colon without fatty separation line. On the sections passing through the thoracic level: right pleural effusion of medium abundance with pulmonary parenchymal condensation opposite.

The surgical exploration by laparotomy found a huge mass with exocystic vegetations, very vascularized, bleeding on contact, coming into contact with the uterus which is visible only at its bottom, adherent to the intestines with peritoneal carcinosis and ascites of great abundance. A tumor reduction was performed with puncture of the cystic content, multiple epiploic and peritoneal biopsies. The anatomopathological examination concluded to a carcinosarcoma of the ovary with epiploic and peritoneal invasion classified IIIC, patient candidate to a carbo pacli based chemotherapy then evaluation.

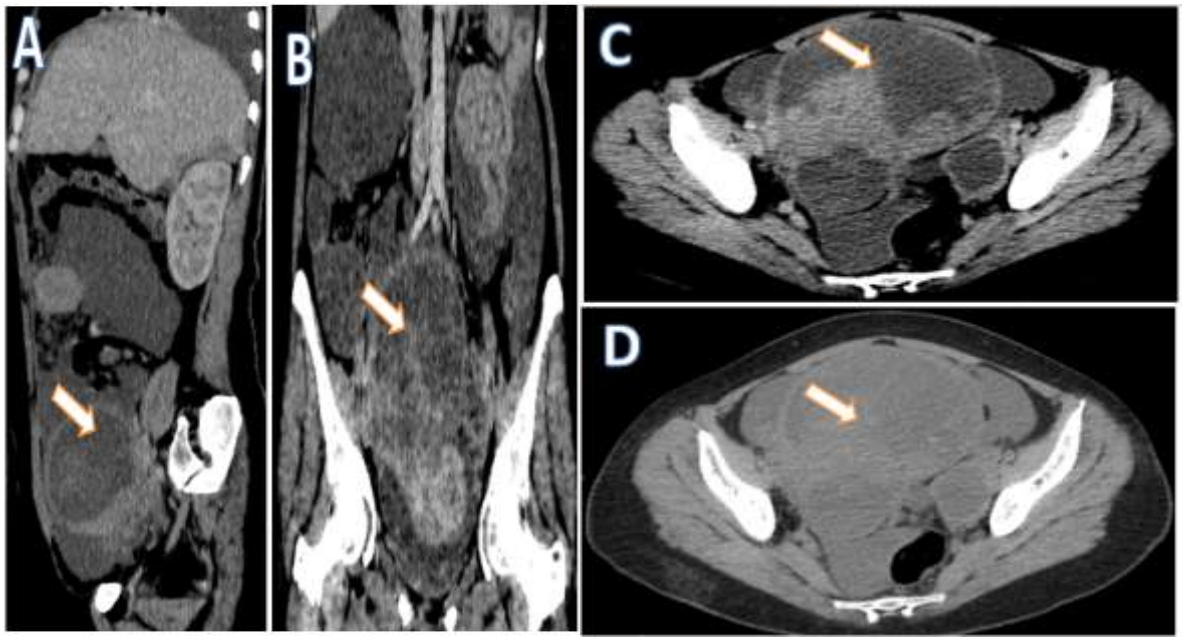

Figure 1(Mme N.B) : Thoracic-abdominal-pelvic CT C-/C+ :Large heterogeneous abdominal-pelvic mass with predominantly suprasternal tissue (arrow) A A:sagittal section B :coronal section C,D :axial sections with and without contrast injection

Discussion:

The term "carcinosarcoma" was first used by Virchow in 1864 to describe a tumor with a carcinomatous and a sarcomatous component [1]. It is a rare tumor that occurs in the female reproductive tract, most commonly in the corpus uteri and less commonly in the cervix, vagina, fallopian tubes or ovary [2]. CSO affects women, most often nulliparous, between the sixth and seventh decade. The age at diagnosis is significantly higher in OSC than in epithelial ovarian tumor (EOT) [3-6]. Depending on the sarcomatous component, two types are defined: either the sarcomatous component is normally present within the organ, in which case it is referred to as homologous carcinosarcoma, or the component is made up of elements that are usually absent (cartilage tissue, bone, striated muscle fibers, etc.), in which case it is referred to as heterologous carcinosarcoma (the most frequent form) [7]. The tumor composition may vary during the course of the disease: metastatic sites are epithelial in about 70% of cases, mixed (epithelial and sarcomatous) in 25% of cases, and with a simple sarcomatous component in 5% of cases, carcinomatous elements are in the majority, while in case of recurrence, sarcomatous elements predominate [8-10].

The clinical presentation is very similar to that of ovarian and uterine carcinomas, with no particular specificity. Clinical and radiological evaluation frequently underestimates the extent of the disease. Very often, the diagnosis is made at an advanced stage of the disease [2,6,8,10]. The metastatic locations are also not different from those of epithelial tumors of the ovary [10]. Rustin and Brown studied the value of CA 125 in OSC [6,4,5]. It is increased in 75-85% of cases [6,10]. Although not validated, it seems to be an interesting marker for therapeutic evaluation in the absence of clinical or radiological criteria. Carcinosarcoma is an aggressive tumor, with a median survival of 7 to 27 months.

The initial stage is the only prognostic factor found in the different studies [8,11,12]. The more advanced the stage, the worse the prognosis in our patient's case. The size, the histological type (heterologous or homologous), the age, are not involved in the prognosis.

Because of the rarity of CSO, there is no consensus on its management. There is very little data. The essential role of surgery seems to be well established, and it is recommended that "ovarian-type" surgery be performed, i.e. total hysterectomy with bilateral adnexectomy, omentectomy, pelvic and lomboaortic curages and peritoneal exeresis with a view to complete resection. Brown, in his comparative study, showed significantly that optimal surgery resulted in longer median survival (14.8 months for optimal surgery versus 3.1 months for suboptimal surgery for stage III, $p = 0.0003$) [12]. [12].

For adjuvant therapy, the only published trial is that of Tate Thigpen for the Gynecologic Oncology Group [13]. The substance used was cisplatin. The response rate is 20%, which is comparable with that observed in uterine carcinosarcoma. Given the low

incidence of OSC, the establishment of therapeutic trials is difficult. Ifosfamide and cisplatin are, then, the two most interesting substances (doxorubicin having shown less efficacy in a GOG study on uterine sarcomas) [13,14,15].

CONCLUSION :

Given its aggressive nature and poor prognosis, OCS requires treatment by radical surgical resection and careful follow-up by CT scan (14). Very few cases have been reported in the literature with frequent recurrences, which makes it difficult to adapt a Consensus for its management.

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