

Clear Cell Adenocarcinoma of Ovary: About a Rare Case and Review of the Literature

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Abstract :- Clear cell adenocarcinoma of ovary is a rare tumor and represents less than 5% of all malignant ovarian tumors. It is an entity distinct from epithelial carcinoma of the ovary and may result from malignant transformation of endometriosis, usually unilateral and highly aggressive. Surgery and chemotherapy are therapeutic weapons even if this tumor has a poor prognosis. We are reporting a case of clear cell adenocarcinoma of ovary, on a young pauciparous patient whose initial therapeutic treatment was effective.

Keywords: Clear cell adenocarcinoma, Endometriosis, Surgery, Prognosis.

I. INTRODUCTION

Clear cell ovarian adenocarcinoma is a rare and extremely aggressive tumor. It could be confused with other ovarian tumors such as: vitelline tumor, ovarian goiter, endometrial carcinoma or even to ovarian metastases from clear cell renal carcinoma. Special coloring and immunohistochemical tests are of great help for the correct diagnosis. It is thought to be the result of the malignant transformation of an endometriotic lesion or a clear cell fibroadenoma [1]. It is a tumor that is usually resistant to first-line chemotherapy for ovarian cancer [2].

II. OBSERVATION

The subject here was a 44-year-old pauciparous patient with no particular medical background other than a secondary dysmenorrhea. She was admitted in a dyspnea chart on a chronic voluminous abdominal mass initially painless. The patient presented no sign of metrorrhagia, fever, nor other associated signs. Clinical examination projected an abdominopelvic mass extending beyond the umbilicus associated with dullness and left pleural effusion syndrome.

The thoraco-Abdomino-pelvic scanner and pelvic MRI revealed a large solidocystic tumor mass, right latero-uterine measuring 19 x 15 cm, probably of ovarian origin with peritoneal carcinoma. A large abundance of left pleurisy was associated (pictures 1, 2, 3). with respect to the emergency the pleurisy was drained, and a pleural biopsy was performed. The histological study concluded that there was no malignancy.

Secondly, with the impossibility of performing an ultrasound-guided tumor biopsy due to the inaccessibility of the carcinoma, and due to the unavailability of laparoscopic surgery, it was decided and performed an exploratory laparotomy which revealed: an ascites of large abundance (about 3 liters), was aspirated while carrying out a cytology; We have noted the presence of a right solidocystic ovarian tumor measuring 20 cm by 18 cm, accidentally ruptured; with peritoneal carcinoma disseminated throughout the abdomen. A Right lumbar aortic lymphadenopathy of 1 cm as well as bilateral centimetric iliac lymphadenopathy were palpable. The rest of the exploration was with no particularity. A right adnexectomy, multiple biopsies of the peritoneum associated with the enucleation of the carcinoma nodules were performed.

The anatomopathological examination of the samples had concluded that there was a well-limited and well-encapsulated carcinomatous proliferation at the ovarian level, with papillary and tubular architecture. Hemorrhagic areas were noted, atypia marked by hyperchromatic and nucleolate nuclei, with loss of nuclear polarity giving the appearance of "upholstery nail". Their cytoplasm was eosinophilic. It was also found microcalcifications and large foci of necrosis. It was therefore a histological appearance suggesting a clear cell adenocarcinoma of the right ovary. The histology of the biopsies on the left ovary concluded in inflammatory changes. Analysis of ascites fluid was non-contributory. The patient received adjuvant chemotherapy with seven cure of Gemcitabine and a treatment with Avastin. A Good disease control has been observed.

III. DISCUSSION

The incidence of ovarian clear cell ovarian adenocarcinoma remains variable in women living in the United States. It is 4.8% for white Caucasians, 3.1% for blacks and 11.1% for Asians. In Japan, the prevalence of ovarian clear cell adenocarcinoma appears to be higher than in Western countries. Its incidence is estimated at 25% of epithelial ovarian cancers [3]; and it accounts for 10% of ovarian cancers [4]. The Clear cell adenocarcinoma carcinogenesis is thought to result from a malignant transformation of endometriosis lesions. Gene mutations and ovarian microenvironmental influence could explain this malignant transformation [1,4].

Our patient reported dysmenorrheic cycles which had never been investigated. Clinically, there is no specificity for clear cell adenocarcinoma; patients generally consult late for an abdomino-pelvic mass with or without compressive signs and/or distant manifestations such as pleurisy. This was the case with our patient. When it comes to imagery, there are no direct signs. On the other hand, characteristic images were found suggesting a highly suspicious lesion. Bouic-Pages in a work had established a concordance between the MRI result and the macroscopic appearance of clear cell adenocarcinoma of ovary. He described a unilocular cystic lesion with rounded solid protrusion taking the contrast [5]. This aspect could already make consider the continuation of the assumption of responsibility which resolutely passes by the surgery.

Indeed, surgery is the first treatment for this tumor. It consists of a total hysterectomy without conservation of the appendages, with omentectomy and lymphadenectomy to the lumbar aortic nodes. In this case, we did not perform a lymphadenectomy. However, it is recognized that patients who have undergone lymphadenectomy have longer progression-free survival than those who have not received it at all stages. Clear cell adenocarcinoma is a tumor generally resistant to platinum salts and taxanes (standard treatment for epithelial tumors of the ovaries). Clinical trials using targeted therapies are currently being evaluated and show promise [2].

Another important problem in the clinical management of clear cell adenocarcinoma of ovary is the lack of effective chemotherapy against recurrent disease after first-line treatment [3]. There are no specific tumor markers in the malignant transformation of endometriotic lesions [6]. In some studies, the evolution of malignant tumors from endometriosis appears to have a relatively good prognosis with five-year survival rates of 65% for ovarian lesions and 100% for extra-ovarian lesions [7]. Our patient has benefited from 7 cure of chemotherapy with good clinical and radiological control even if we do not have sufficient hindsight.

IV. CONCLUSION

Clear cell adenocarcinoma of the ovary is a rare tumor that can have a poor prognosis. The Targeted therapy thanks to advances in molecular biology offers hope for the management of this tumor.

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Fig 1: Thoracic level CT scan showing left pleural effusion.

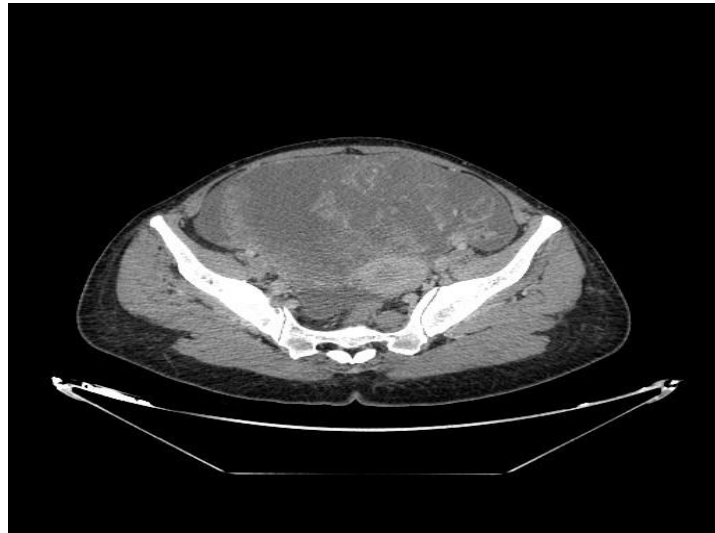


Fig 2: Pelvic floor CT scan showing a right latero-uterine mass.

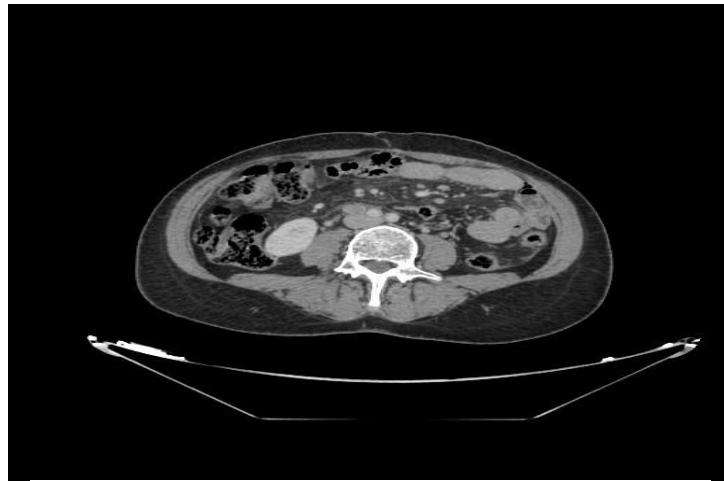


Fig 3: Abdominal stage CT scan showing peritoneal carcinoma.