Hormone-Sensitive Primary Peritoneal Sammocarcinoma: A Case Report and Literature Review

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Abstract

Psammocarcinoma is a rare low-grade serous carcinoma resulting from an ovary or peritoneum. It is characterized by its richness in psammomatous calcospherites and an invasion of surrounding structures. Its prognosis is much more favourable than serous carcinoma of the ovaries and peritoneum. We report a new case of serous peritoneal psammocarcinoma in a 50-year-old patient who was treated with Anastrozol and LHRH analog hormone therapy for one year. The clinical and radiological response was complete. The surgery was carcinological and consisted of a total hysterectomy with adnexectomy and omentectomy.

Keywords: borderline serous tumor, peritoneum, psammocarcinoma, hormone therapy.

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INTRODUCTION

Psammocarcinoma is a rare type of serous carcinoma from the ovaries or peritoneum. Primary Peritoneal Psammocarcinoma (PPP) is characterized by dystrophic calcifications called psammoma bodies, with minimal or no damage to the ovaries and invasion of surrounding structures and low-grade cytological atypia[1]. The clinical behaviour of the PPP is variable. The majority of them have a relatively favourable prognosis and some PPPs may have recurrences and metastases [2, 3].

In this work, we will present a rare case of PPP that has been confirmed by histology and immunohistochemistry. It was treated with hormone therapy and surgery.

CASE REPORT

A 50-year-old patient with no particular pathological history had isolated pelvic pain in March 2006. The clinical examination was without particularity. A thoraco-abdomino-pelvic CT scan of the patient showed a pure cystic mass of the right ovary of 61×84 mm² with a regular thin-walled contour, calcified at its anterolateral external part. These calcifications extend intraperitoneally by contiguity. The CA 125 tumor marker was 81 U/ml.

The patient had a right oophorectomy with a peritoneal biopsy and sampling of the peritoneal fluid. The anatomo-pathological study concluded that there was a fibro-inflammatory reworking of the ovarian parenchyma and dystrophic and inflammatory calcifications of the peritoneum. Two years later, the patient had a recurrence of abdominopelvic pain. The clinical examination had revealed a mass on the left flank and periumbilical. The CA125 was 61U/ml.

The MRI had showed an intraperitoneal mass of the left flank that extends towards the poorly individualized contralateral infiltrating side measuring 16.4×4.5 cm². This mass does not invade the abdominal muscles (Figure 1 and 2). Surgical exploration had revealed a mass in the left flank and iliac fossa bleeding on contact whose carcinological surgical excision was impossible.
The T1 axial plane image with Gadolinium injection shows an intra-abdominal mass on the infiltrating and poorly individualized left flank, measuring 16 × 3.5 cm² on the axial plane and 16 × 4.3 cm² on the sagittal plane. It is in contact with the abdominal muscles that invade and extends to the white line without lymphadenopathy or ascites.

The anatomopathological study of the tumor biopsies identified infiltration of parietal tissues by epithelial malignant tumor proliferation rich in psammomas and tumor cells with mild atypia. The stroma-reaction was grouped into nodules. The Immunohistochemical study confirmed the diagnosis of psammocarcinoma by detecting diffuse positivity of proliferative cells to EMA, CK7, AE1, AE3 and hormone receptors to estrogens and progesterones as well as focal positivity of proliferative cells to WT1. The marking with CK5/6 CK20 CD2 and catreitin was negative.

The patient was started on LHRH analog hormone therapy, 3.6 mg monthly and Anastrozole 1mg daily. The 3-month evaluation of CT treatment showed a reduction in tumor volume of more than 50% (Figure 3 and 4).

The CA125=24U/ml after one year of hormone therapy, abdominopelvic MRI had no residual tumor and CA125 normalized to 11.1 U/ml (Figure 5 and 6). The patient had a total hysterectomy with appendectomy, omentectomy and appendectomy. The annexes, appendix and peritoneum are the site of parietal calcospherites without associated epithelial or mesothelial proliferation. At the time of writing this article, the patient is in good control with a 4-month delay after the surgery.
Fig-5 and 6: T1 and T2 images with Gadolinium injection show the persistence of an infiltrating process of the heterogeneous left flank which is poorly limited by 140 × 70 mm2 without any focus of hyper signal or restriction on the diffusion sequences nor any enhancement after injection of Galonium.

DISCUSSION

The epithelium of the ovary and the mesothelium of the peritoneum have the same embryological origin, but PPP is rarer than that of the ovary. To date, about 30 cases of peritoneal and ovarian psammocarcinoma have been reported in the literature [3-9].

In clinical terms, PPPs are usually asymptomatic or sometimes associated with non-specific symptoms such as abdominal pain, nausea, vomiting, and heavy menstrual bleeding [4,5]. PPP can be detected accidentally by increasing CA-125[2-4]. Aggressive forms are usually associated with high CA-125 values. The average age of the patients is 54 years (extreme, 27-84 years)[4,6]. The patient in our study was 50 years old.

PPP is histologically identical to epithelial ovarian carcinoma. The main differentiation characteristic is the advanced peritoneal involvement of the tumor without coarse involvement of the ovaries [3]. The following pathological characterizations for peritoneal psammocarcinoma have been reported by Gilks et al.[10]

- Destruction of the intraperitoneal stroma.
- Nuclear atypia no more than moderate.
- No solid epithelial proliferation zones except occasional nests ≤ 15 cells in diameter.
- At least 75% of the papillaries or associated nests are completely replaced by psammoma bodies.

Chen et al. extended these criteria to qualify psammocarcinoma as peritoneal psammocarcinoma, including either the invasion of intraperitoneal viscera or invasive nodules into the peritoneum [11]. The mechanism of appearance of psammomas is not yet confirmed but there is a dystrophic calcification of the cell by the accumulation of hydroxyapatite in degenerating cells replaced by psammoma bodies [2-4].

Given the scarcity of PPPs, there are no standard protocols. Total hysterectomy with adnexectomy and omentectomy are recommended for optimal debulking[3,6,10]. Conservative surgery can be done for young women with PPP without macroscopic damage to both ovaries in order to preserve their fertility [3]. Since PPP is a low-grade, low mitotic index serous carcinoma, adjuvant or neo-adjuvant chemotherapy is not very effective. However, it may be considered in patients with residual peritoneal disease after surgery or for PPP with aggressive behavior [3,10].

To my knowledge, only a few cases of PPP have been treated with hormone therapy in the literature [9]. For our patient, the estrogen and progesterone hormone receptors were positive. Hormone therapy with Anastrozole and LHRH Analog over 12 months resulted in a complete radiological and biological response and optimal carcinological surgery. On the excised tissues, there was a minimal microscopic residue.

CONCLUSION

The treatment of peritoneal psammocarcinoma consists of debulking surgery. Adjuvant chemotherapy did not show an increase in survival. Hormone therapy may provide a complete response in cases where hormone receptors are present on tumor cells.

REFERENCES