

Primary ovarian serous psammocarcinoma-a case report with mini literature review

Abstract

Serous psammocarcinoma is a rare subtype of serous carcinoma in which a significant number of psammoma bodies are present histologically. Because it is an extremely rare disease, the pathogenesis of the disease remains unclear. Here we present a case of primary ovarian serous psammocarcinoma in which computed tomography (CT) imaging of the abdomen was useful for the diagnosis.

A female patient was referred to our department with a complaint of abdominal distention. Pelvic magnetic resonance imaging revealed a multifocal tumor with a papillary enhancement in the right appendicular region, and abdominal CT imaging exhibited marked calcification of the enhancement. She underwent a laparotomy for suspected right ovarian cancer. As postoperative pathological examination revealed dominant stromal infiltration of psammoma bodies beneath peritoneum, this case was diagnosed as primary ovarian serous psammocarcinoma at FIGO Stage IIB. The patient was treated with six courses of paclitaxel plus carboplatin after the surgery. She is currently under outpatient observation without any signs of recurrence after 8 years of treatment. The case of significant calcification of the intra cystic enrichment on CT imaging is considered to be a case of this disease.

Keywords: psammocarcinoma, serous carcinoma, ovary, peritoneum, psammoma body

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Introduction

Serous psammocarcinoma is a very rare disease classified as one entity of ovarian serous carcinoma. Many psammoma bodies are found in tumor tissue, and their prognosis is relatively good.^{1,2} We report a case of primary ovarian serous psammocarcinoma in which preoperative computed tomography (CT) examination of the abdomen was useful for diagnosis.

Case report

The patient presented here is 59 years old female with 1 para. She was menopausal at 50 years old. There was no remarkable episode in previous medical or family history. She presented to the previous doctor for abdominal distention. Ultrasonography revealed an abdominal tumor, and the patient was referred to our department for further examination and treatment. At the first visit, a mass reaching umbilical height in the lower abdomen was palpated. On bicuspid examination, the mass was supernumerary head size and its mobility was good. Serum CA19-9 and serum CA125 levels were as high as 1987 and 1861U/ml, respectively. Pelvic magnetic resonance imaging (MRI) revealed a 14x10 cm multifocal tumor in the right ovary and a papillary-like enhanced component within the cyst that showed iso to high signal on T2-weighted images (Figure 1). Abdominal computed tomography (CT) revealed a solid component with marked calcification and contrast effect in the tumor cyst (Figure 2). FDG PET-CT also showed high signal accumulation on the solid component in the tumor cyst (Figure 3).

She underwent abdominal simple hysterectomy, bilateral salpingo-oophorectomy, partial omentectomy, and pelvic and par-aortic lymph node dissection for suspected right ovarian cancer. There was a small amount of serous ascites in the abdominal cavity. The right adnexae was enlarged to the size of the supernumerary head, but there were no adhesions to the surrounding tissues. The multilobulated intratumoral solution was serous, and the segment of the enhancement

was yellowish and rough (Figure 4). Tumor-printing cytology showed round, similarly shaped tumor cells forming papillary-like masses on a relatively clear background. The nuclei were relatively homogeneous and round, and the nuclear/cytoplasm ratio was low. The sand grain bodies were not distinct. Based on these findings, low grade serous carcinoma was suspected (Figure 5). Postoperative pathological examination revealed that monolayered cubic tumor cells with mild to moderate cellular atypia proliferated in the architecture of glandular tubules or papillae with stromal infiltration. In addition, tumor stroma was dominantly infiltrated with sand grains small bodies (Figure 6). Very focal necrosis was observed in the tumors. We found non-invasive disseminated implantation the vesico-uterine fossa, but no dissemination or metastasis in other organs or lymph nodes (pT2bN0M0). On immunohistochemical staining, tumor cells were positive for WT-1 and ER, but negative for p53 (Figure 7). Based on the above findings, the diagnosis of primary ovarian serous psammocarcinoma was made. Postoperatively, six courses of paclitaxel (175mg/m²)/carboplatin (AUC 6) were administered. She is currently under outpatient observation without any signs of recurrence after 8 years of treatment.

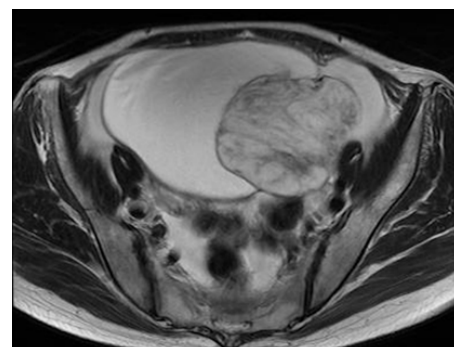


Figure 1 Pelvic contrast MRI. T2-weighted images showed a 14-cm-diameter polycystic tumor in the pelvis. In the cyst, there was a solid component that showed iso to high signal.

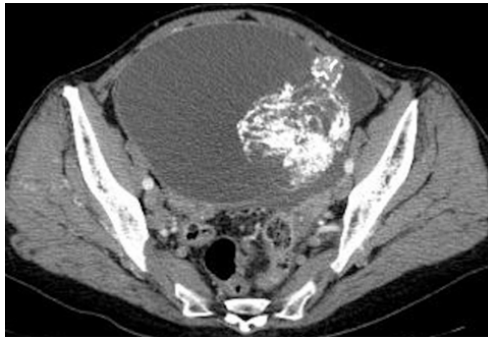


Figure 2 Abdominal contrast CT. The cyst was markedly calcified and had a solid component with contrast effect.

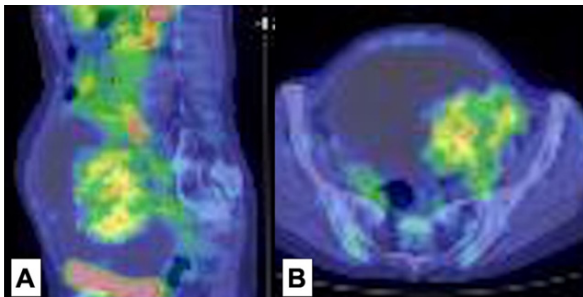


Figure 3 PET-CT. FDG signal accumulation on the enhanced area in the tumor cyst. A. Sagittal section, B. Transverse section.

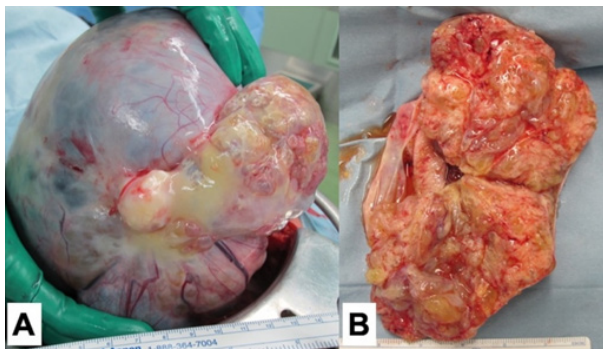


Figure 4 Surgical excision specimen.

- A. Tumor enlarged to 14 cm in size, consisting of a cyst and a substantial portion.
- B. Grossly visible, the tumor is white to pale yellow in color; and there is a congestion of vesicles.

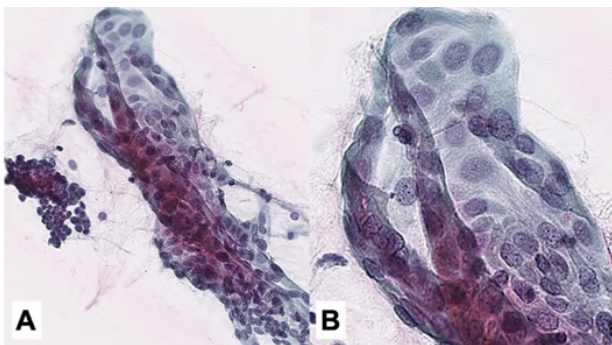


Figure 5 Tumor-printing cytology (Papanicolaou staining). Tumor cells of similar round shape appear in papillary-like masses against a relatively clear background. The nuclei were relatively homogeneous and round, and the N/C ratio was low. A. $\times 100$, B. $\times 400$.

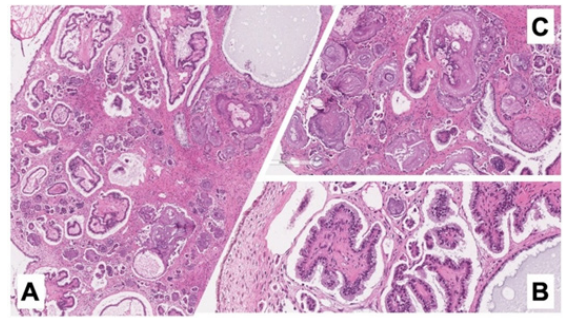


Figure 6 Histopathological examination (Hematoxylin Eosin staining).

- A. Monolayered cubic tumor cells with mild to moderate cellular atypia proliferated in the form of glandular tubules or papillae with interstitial infiltration. ($\times 100$).
- B. Monolayer cubic tumor cells showing mild to moderate cell atypia. ($\times 200$).
- C. A psammoma body was observed in the stroma, occupying more than 75 % of the total. ($\times 100$).

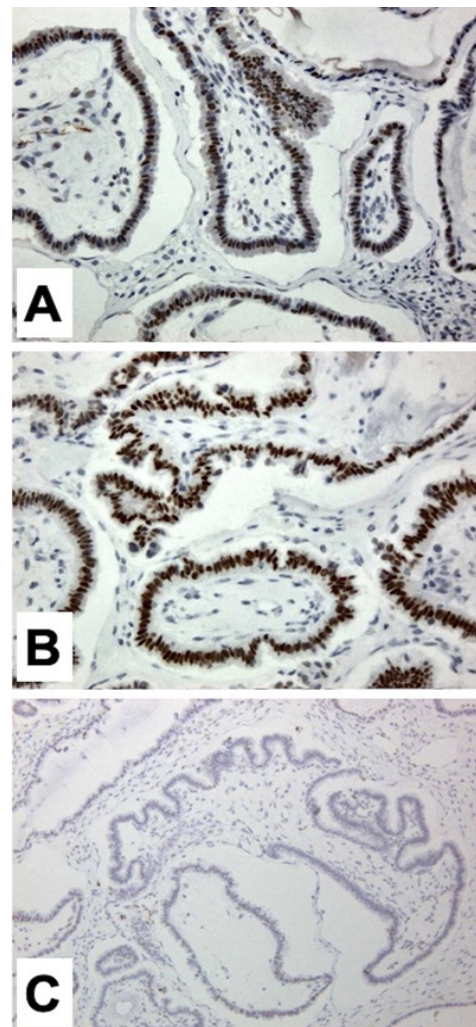


Figure 7 Immunohistochemical examination.

- A. Immunostaining for WT-1. Positive image of atypical cells lining the tumor wall. ($\times 200$).
- B. Immunostaining for ER. Positive image of atypical cells lining the tumor wall. ($\times 200$).
- C. Immunostaining for p53. Both atypical cells and tumor stroma are negative. ($\times 100$).

Discussion

Psammocarcinoma of ovary was first reported by Kettle et al. in 1916 as a serous carcinoma containing a large amount of sand grains³, and since then there have been various reports on its clinical concept and characteristics. Sand granules are stratified calcifications formed by deposition of calcium phosphate on tumor cells and are commonly found in papillary thyroid carcinoma, meningioma, and serous ovarian carcinoma. Ovarian primary psammocarcinoma has been classified as a subtype of serous carcinoma with a relatively good prognosis.^{1,2} On the other hand, it has been reported that the presence and extent of sand grains do not correlate with prognosis.^{4,5}

On imaging examination, although CT scan exhibits a large amount of calcification in the ovarian tumor, it is very difficult to rule it out from other benign calcified diseases such as calcified leiomyoma, Brenner tumor, and fibrothecoma. On the other hand, there is a report that FDG PET/CT may be useful in differentiating malignancy.⁶ This case was suspected of having this disease because of the contrast CT examination showing the contrast effect with marked calcification in the enhanced area. In addition, FDG PET/CT showed abnormal accumulation in the same area, which made it relatively easy to designate this case as malignant.

In Psammocarcinoma, it has been reported that small sand grains, psammoma bodies, were detected by preoperative cervical cytology, ascites cytology or intraoperative abdominal washing cytology, and tumor imprinting cytology.⁷⁻⁹ Although no sand granules were found on any cytological specimen in this case, tumor-imprinting cytology exhibited the cell clusters of low grade serous carcinoma, which may help in the diagnosis of psammocarcinoma. Gilks et al. proposed four criteria for histopathological diagnosis:

- a) Invasive carcinoma
- b) No more than moderate cell atypia
- c) No enriched proliferation of cancer cells with more than 15 cells in clusters
- d) Presence of sand granules in more than 75% of papillary growth foci.¹

The present case also met all four of these criteria and was diagnosed as psammocarcinoma. In immunohistochemistry, tumor cells are positive for CK7 and ER and negative for CK20, calretinin, and Desmin.¹⁰ In the present case, tumor cells were positive for ER and WT-1 (Figure 7).

After reviewing previous reports, we found that 23 out of 26 patients bearing psammocarcinoma of ovary were advanced at FIGO stage III-IV (88%), which indicates a trend similar to that of ovarian high-grade serous carcinoma.^{1,10-22} The survival rate is higher than that of high-grade serous carcinoma and is comparable to that of borderline malignant serous tumors, and the 5-year survival rate of patients at stage I has been reported to exceed 95%.^{23,24} Even in advanced cases, the rate of recurrence is low, and in cases of recurrence, the progression is gradual and follows a long-term course.^{1,25} Although there is no standard treatment for this disease due to the rarity, intensive cytoreduction and postoperative adjuvant chemotherapy are recommended as the initial treatment.^{20,26} However, the frequency of recurrence has been reported to be as high as 70%, and there have been many cases which repeated recurrence and died of the disease.²⁶⁻²⁸ Although the efficacy of chemotherapy is not clear, there have been reports of relapse in patients who did not receive

adjuvant chemotherapy.^{18,29} In this case, adjuvant chemotherapy was administered postoperatively and a good prognosis was obtained, suggesting a certain efficacy of chemotherapy.

In this study, we experienced a case of primary ovarian serous psammocarcinoma in which preoperative abdominal computed tomography was useful for the diagnosis. It is mandatory to establish the strategy of early diagnosis and treatment for this rare tumor by the accumulation of cases.

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Conflicts of interest

All authors declare that they have no competing interests.

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