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Small cell ovarian carcinoma of the pulmonary type: a rare case

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Summary

Background: Small cell ovarian carcinoma of the pulmonary type is extremely rare. It is extremely aggressive and progressive, and there are no established treatment guidelines. *Case:* A 55-year-old woman presented with small cell ovarian carcinoma of the pulmonary type. Postoperatively the patient received chemotherapy combining cisplatin and irinotecan. To date, nine months after operation, the patient is alive with no evidence of recurrence or metastasis. *Conclusion:* In case of small cell ovarian carcinoma of the pulmonary type, chemotherapy regimen used for small cell lung cancer, such as chemotherapy combining cisplatin and irinotecan, should be considered.

Key words: Ovarian cancer; Small cell carcinoma of the pulmonary type.

Introduction

Small cell ovarian carcinoma is a rare type of ovarian cancer, and divided histologically into the hypercalcemic type and the pulmonary type [1-3]. Especially, the pulmonary type is extremely rare [1]. It is extremely aggressive and progressive, and there are no established treatment guidelines for this tumor type [1-3]. Previous reports concluded that treatment of small cell ovarian carcinoma of the pulmonary type should be based on the therapies used to treat other small cell carcinomas [1,3]. Here, we present a rare case of small cell ovarian carcinoma of the pulmonary type.

Case Report

A 55-year-old woman without a past medical history of interest was referred to the present hospital with atypical genital bleeding. No other symptoms were observed. Ultrasonography revealed a large pelvic tumor without ascites. Magnetic resonance imaging (MRI) examination of the pelvis, which demonstrated a large left adnexal mass with an internal focus of bleeding, suggesting an ovarian cancer (Figure 1). Computed tomography (CT) examination of the abdomen and chest demonstrated no metastasis. Laboratory investigations showed no remarkable findings including serum Ca level. Serum tumor markers were as follows: CA125: 119 U/ml, CEA: 0.7 ng/ml, CA19-9: 2 U/ml, STN: 17.6 U/ml, and neuron-specific enolase (NSE): 6.8 ng/ml. The cytological examination of cervix and the histological examination of endometrium showed no abnormality. Therefore, primary ovarian cancer was initially considered the most likely diagnosis.

The patient underwent an exploratory laparotomy. Macroscopically, there were a moderate amount of bloody serous ascites in the peritoneal cavity. There was left ovarian tumor (size: 15×13 cm) which was yellow-white in color and solid with necrotic lesions. The right ovary and uterus were unremarkable without a small uterine myoma (Figure 2). There were no other remarkable findings in the peritoneal cavity. The frozen pathological examination of the resected specimen showed poorly differentiated carcinoma of left ovary, including a dense proliferation of cellular oval cells with scanty cytoplasm. At this time, the authors diagnosed the patient as primary ovarian cancer and performed hysterectomy, bilateral salpingo-oophorectomy, partial omentectomy, and pelvic lymph node biopsy. For massive adhesion and bleeding, systematic pelvic lymphadenectomy and para-aortic lymphadenectomy were not performed.

The pathological examination showed that the tumor was

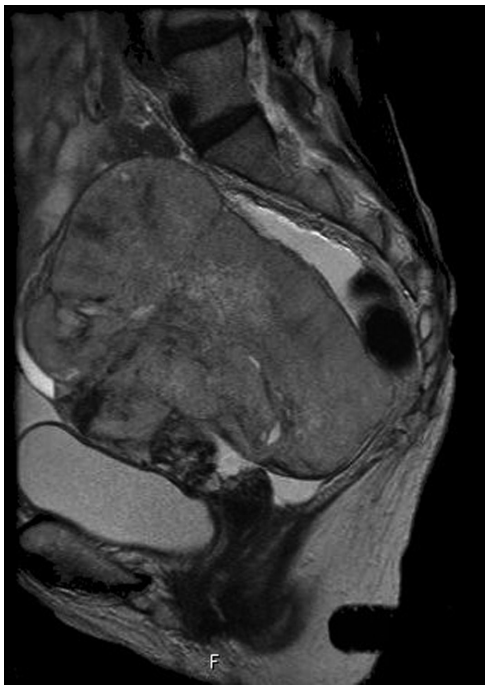

Figure 1. — MRI examination of the pelvis which demonstrates a large left adnexal mass with an internal focus of bleeding. There are few ascites and no evidence of tumor invasion to the surrounding tissue.

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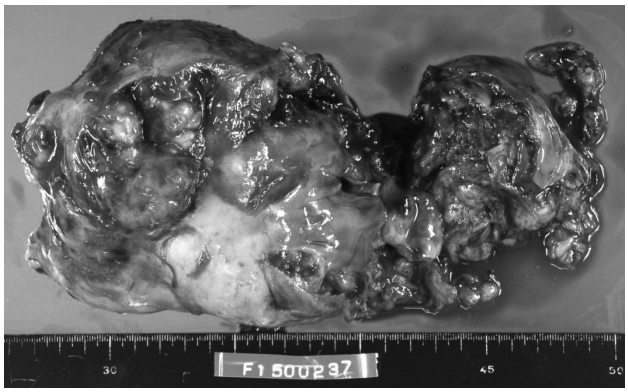

Figure 2. — Macroscopic image of resected specimen. There is left ovarian tumor (15×13 cm) which is yellow-white in color and solid with necrotic lesions.

densely cellular, and the tumor cells had scanty cytoplasm and small to medium-sized hyperchromatic nuclei that were oval to spindle shaped. The tumor cells showed strong positivity by immunohistochemistry for cluster of differentiation 56 (CD56) and synaptopodin, although chromogranin A was negative (Figure 3). Finally, the pathological examination showed a small cell carcinoma of the pulmonary type. The authors confirmed the diagnosis of primary ovarian cancer Stage IC1 (pT1cNXM0) of left ovary according to the International Federation of Gynecology and Obstetrics (FIGO) surgical staging system.

At two weeks after operation, the patient received six courses of chemotherapy combining cisplatin (60 mg/m^2, Day 1), and irinotecan (60 mg/m^2, Days 1, 8, and 15) administered every four weeks. To date, nine months after operation, the patient is alive with no evidence of recurrence or metastasis.

Discussion

Small cell ovarian carcinoma of the pulmonary type is extremely rare [1-3]. To the present authors' knowledge, there are only 36 reported cases of small cell ovarian carcinoma of the pulmonary type [3, 4]. It resembles small cell carcinoma of the lung [1-3]. Eichhorn *et al.* reported 11 patients with small cell ovarian carcinoma of the pulmonary type. In the long-term follow-up, they died of the disease at one to 13 (mean eight) months [3]. Munstedt *et al.* reported 34 patients with small cell ovarian carcinoma of the pulmonary type. It has a poor prognosis even when diagnosed at an early stage, and radical surgery was not as successful as in the treatment of common epithelial ovarian carcinomas [1].

Previous reports conclude that treatment of small cell ovarian carcinoma of the pulmonary type should be based on the therapy used to treat other small cell carcinomas [1-3]. Munstedt *et al.* report that 82.4% (28/34) of patients received additional postoperative chemotherapy, and chemotherapeutic regimens mainly consisted of cisplatin or carboplatin (83.9%), etoposide (41.9%), alkylating agents (cyclophosphamide or ifosfamide) (25.8%), pacli-

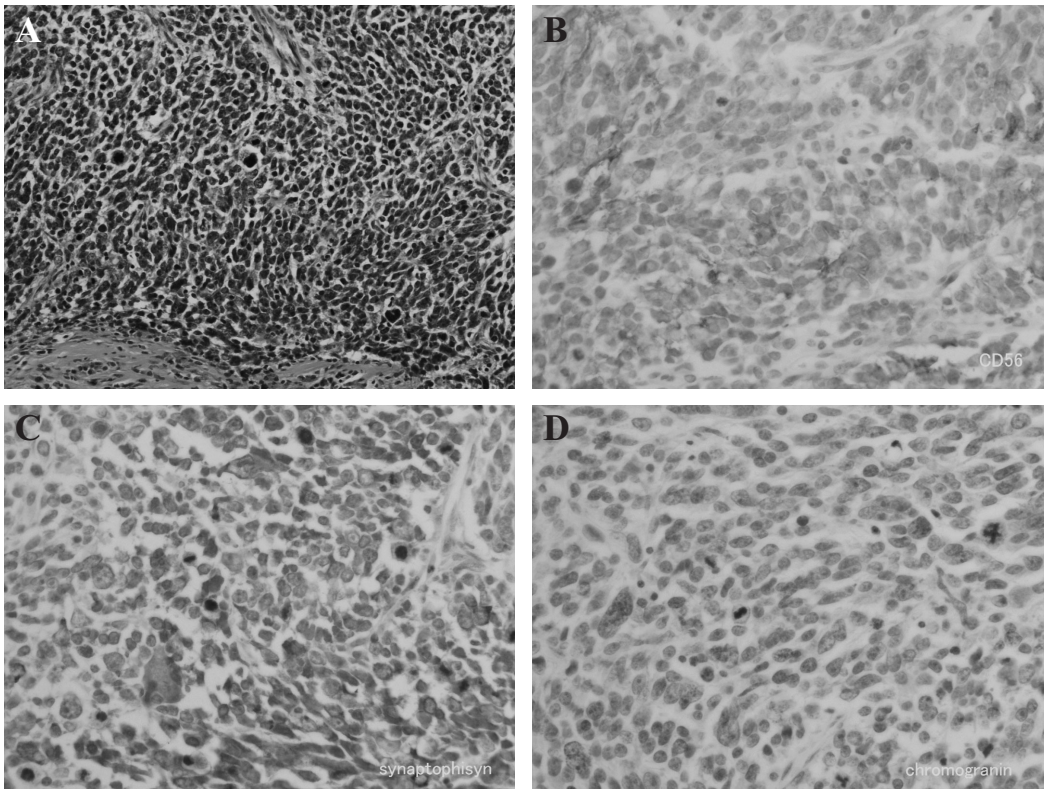

Figure 3. — A. The microscopic image of tumor cells (Hematoxylin and eosin, ×20), which showed scanty cytoplasm and small to medium-sized hyperchromatic nuclei. B. The image of CD56-positive tumor cells (×40). C. The image of synaptopodin-positive tumor cells (×40). D. The image of chromogranin A-negative tumor cells (×40).

taxel (19.4%), doxorubicin (12.9%), bleomycin (12.9%), vinca alkaloids (6.5%), and irinotecan (6.5%). They found a trend towards improved survival with the use of etoposide and anthracyclines [1]. In the present case, the authors chose a chemotherapeutic regimen including irinotecan and cisplatin, which are currently used for small cell lung cancer and are considered to be effective for small cell carcinoma of the ovary. There is only one report using chemotherapy combining cisplatin and irinotecan. In that case, the patient diagnosed with ovarian carcinoma classified as Stage IIIc, was treated with chemotherapy combining irinotecan and cisplatin after an optimal debulking surgery, and after four courses of chemotherapy, multiple liver metastases and Virchow's lymph node metastases were found. The treatment was ineffective, and she died six months after surgery [2]. In the present case, the patient received six courses of chemotherapy combining cisplatin (60 mg/m^2, Day 1), and irinotecan (60 mg/m^2, Days 1, 8, and 15) administered every four weeks. In spite of insufficient operation without systematic pelvic lymphadenectomy and para-aortic lymphadenectomy, the patient is alive with no evidence of recurrence or metastasis nine months after operation. This regimen seems to be effective for patients with small cell ovarian carcinoma of the pulmonary type.

In case of small cell ovarian carcinoma of the pulmonary type, chemotherapy regimen used for small cell lung cancer such as chemotherapy combining cisplatin and irinotecan should be considered.

References

[1] Munstedt K., Estel R., Dreyer T., Kurata A., Benz A.: "Small cell ovarian carcinomas – characterisation of two rare tumors". *Geburtshife Frauenheikd*, 2013, *73*, 698.

[2] Kira N., Takai N., Ishii T., Kai K., Nishida M., Nasu K., *et al.*: "Ovarian small cell carcinoma complicated by carcinomatous meningitis". *Rare Tumors*, 2012, *4*, e26.

[3] Eichhorn J.H., Young R.H., Scully R.E.: "Primary ovarian small cell carcinoma of pulmonary type." *Am. J. Surg. Pathol.*, 1992, *16*, 926.

[4] Rubio A., Schldt M., Chamorro C., Crespo-Lora V., Nogales F.F.: "Ovarian small cell carcinoma of pulmonary type arising in mature cystic teratomas with metastases to the contralateral ovary". *Int. J. Surg. Pathol.*, 2015, *23*, 388.

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