



A Case of Primary Retroperitoneal Serous Adenocarcinoma and Literature Review

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Abstract

Primary retroperitoneal serous adenocarcinoma (PRSA) is an extremely rare clinical entity with only seven cases previously reported. In this article, we described a 58-year-old woman with a solitary tumor of serous adenocarcinoma located in the right side of the Douglas Pouch arising from the retroperitoneum. Although several theories have been proposed, the exact origin of PRSA remains unclear. The most widely accepted theory is caelomic metaplasia. According to the histopathological results of the present case, the concept of caelomic metaplasia has been proved to a certain extent. As far as we know, this may be the first case which provides a direct and potent evidence for this theory and is conducive to explain the histogenesis of PRSA. The patient underwent a treatment modality combining complete surgical resection with platinum-based chemotherapy, which is extremely likely to contribute to an excellent clinical outcome, and we consider that such therapeutic strategy may be the preferred method for the treatment of PRSA.

Introduction

Primary retroperitoneal serous adenocarcinoma (PRSA) is an extremely rare malignancy, about which only seven cases previously reported as a result of a careful search in PubMed database from 1991 to December 2016 [1-7]. All of the reported patients were women ranging in age from 11 to 75 years. Because of the rarity, the histogenesis of PRSA remains unknown, the biological behavior, clinical manifestation and imaging finding of the tumors are various and the diagnostic criteria and therapeutic strategy of this entity are still not unified [2,5-7]. A solitary solid tumor in the Douglas pouch from the retroperitoneum has not been described in previous reports. Here we report a rare case of PRSA and review the relevant literatures.

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Case Presentation

A 58-year-old postmenopausal woman, gravida2, parity1, first visited our Department in July 2014 with a three-week history of alternate stool abnormality. She had no medical history of abdominal surgery or lesion. A tumor 43*33*26 mm in size located behind the cervix was detected by ultrasound transvaginal examination. A pelvic computed tomography (CT) scan showed a low-density solid neof ormation with an irregular and enhanced margin which was adjacent to the rectum in cul-de-sac and multiple enlarged lymph nodes in celiac and pelvic cavity (Figure 1). Gastrointestinal investigations, including gastrointestinal scopy and colonoscopy were normal. Laboratory studies demonstrated elevated serum levels of CA-125, 3942.00 U/ml.

For further work-up, a laparoscopy was performed. We detected a solitary solid-appearing mass measuring 4 cm in diameter approximately in the right side of the Douglas Pouch, which was located in the retroperitoneum and squeezed the rectum to the left (Figure 2). The size and appearance of uterus, bilateral fallopian tubes, ovaries, peritoneum, omentum majus and mesentery were normal. Considering that the tumor was closely attached to the rectum wall (Figure 3), a surgeon was invited to consult to exclude rectal neoplasm. A complete resection of the retroperitoneal tumor was conducted, and the histopathologic examination of a frozen section showed low-differentiated adenocarcinoma. Subsequently, a total hysterectomy, bilateral salpingo-oophorectomy, omentectomy, pelvic and paraaortic lymphadenectomy and biopsy of the peritoneum were performed.

Cytological analysis of peritoneal washing liquid showed no abnormalities. Postoperative histopathology examination of the specimen showed a poorly-differentiated adenocarcinoma involving the tumor. There were no microscopic invasion of the uterus, ovaries and tubes, omentum and lymph nodes. Immunohistochemistry of the mass pathology revealed positive findings for CK,

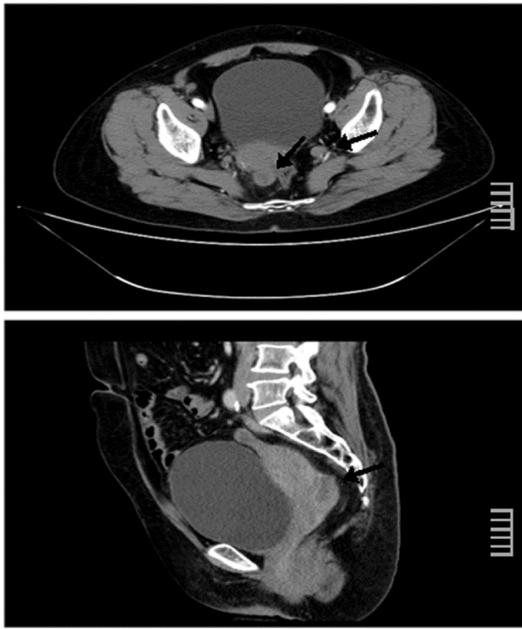


Figure 1: Pelvic computed tomography scan showing a solid tumor in cul-de-sac and enlarged lymph nodes in pelvic cavity (arrows).



Figure 2: A solitary solid-appearing tumor in the right side of the Douglas Pouch from the retroperitoneum.

CA124, and 70% positive for Ki-67, partly positive for MC and negative findings for CEA, Vim, Calret and CDX2. The pathology consultation results in Peking University Third Hospital demonstrated a peritoneum original high-grade serous adenocarcinoma; meanwhile, a small amount of benign mucosal epithelium was seen beside the tumor tissue, which showing morphological differentiation towards to the epithelium of the fallopian tube (Figure 4A and B). Consequently, a histological diagnosis of PRSA was made, and defined at stage II according to the 2006 FIGO classification.

The patient was subsequently treated with platinum-based chemotherapy, a combination of paclitaxel and carboplatin (TC therapy, 3-week intervals). After a total of seven courses of TC therapy, there was a significant reduction on the level of serum CA125 from 2558.00 U/ml to 12.82 U/ml (Figure 5) and CT scan showed no evaluable disease. 21 months after the surgery, positron emission tomographic (PET) scan revealed a retroperitoneal nodule in pelvic cavity and a suspicious bone metastasis in sternum. The patient was once again treated with chemotherapy, which was effective as before. When last seen on January 2017, the patient has been alive with stable disease for 30 months since her surgery and the follow-up is still



Figure 3: The relationship between the tumor and the rectum.

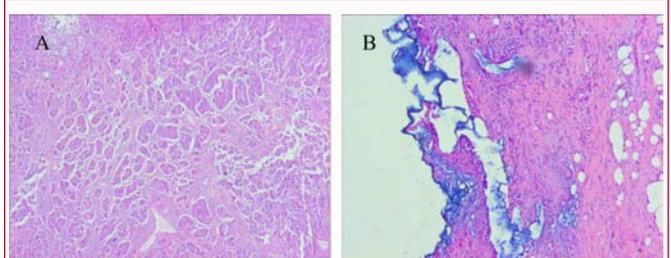


Figure 4: (A) Histological findings of the retroperitoneal tumor, showing solid growth of atypical epithelial cells with pleomorphic nuclei and frequent mitotic figures; (B) a small number of benign mucosal epithelium demonstrating tubal differentiation in morphology beside the tumor tissue (HE staining x100).



Figure 5: Change of serum CA-125 levels along the treatment. **Note:** PREOP: Pre-operation 1: first course of chemotherapy

continuing.

Discussion

PRSA is an extremely rare malignancy disorder with only seven cases previously reported [1-7]. To explain the exact histogenesis of the PRSA, several theories have been postulated including coelomic metaplasia, extraovarian endometriosis, ectopic ovarian tissue, supernumerary ovaries, teratoma and enterogenic cyst, currently the most widely accepted one is caelomic metaplasia. The peritoneum has been defined as the secondary Müllerian system for similar potential to differentiate into various types of epithelium derived from the Müllerian tube: endometrium, endocervix, and fallopian tube [8,9]. It is believed that the epithelial metaplasia of peritoneal mesothelium is one of the originations of the extragenital Müllerian neoplasms. The metaplastic epithelium has a tumorigenic potential, resulting in the formation of a variety of tumors including serous, mucinous, endometrioid and clear cell tumors. Singh et al. [10] point out that there is sufficient and clear evidence to prove that fallopian tube is

the origination of non-uterine high grade serous carcinoma (HGSC). Gruessner et al. [11] have reported a case in which a potential link between endosalpingiosis (ES) and the development of pelvic serous carcinoma in woman has been observed. Therefore, it is conceivable that epithelial retroperitoneal tumors may have initially been metaplastic peritoneal mesothelium.

However, currently there is no case directly confirms that PRSA originates in the metaplasia of peritoneal mesothelium. The pathological consultation result of the present case clearly mentioned that a small number of the benign mucosal epithelium demonstrating tubal differentiation in morphology can be detected beside the tumor tissue, suggesting that the primary retroperitoneal tumor may come from metaplasia of peritoneal mesothelial cells. It confirms that the peritoneum is the matrix where benign and malignant tumors of secondary Müllerian epithelium occur [12]. To our best knowledge, this may be the first case which provides a direct and potent evidence for the theory of caelomic metaplasia and contributes to clarify the histogenesis of PRSA.

It is believed that the biological behavior of PRSA is similar to epithelial ovarian cancer. According to the reported cases, the main clinical symptoms of PRSA are asymptomatic abdominal and pelvic mass, abdominal pain, vomiting, anorexia, abdominal distension and weight loss, resembling those of ovarian serous adenocarcinoma. Despite there is no basic standard of treatment for the disease at present, patients were treated in accordance with the treatment fashion of epithelial ovarian cancer. Among the seven reported cases, four patients underwent complete tumor resection and partial resection of ambient involved tissue. Two of the four patients received chemotherapy, one was alive with disease for 32 months [6] and the other survived 6 months with no clinical evidence of tumor recurrence [7]; the rest two patients did not receive further therapy following surgery, and survived for 24 months [3] and 7 months [5] without recurrence, respectively. Ulbright et al. [1] reported a case of a retroperitoneal neoplasm in an 11-year-old girl. The girl underwent a partial resection of tumor and chemotherapy and was alive with no evidence of disease for 10 months. Fujiwara et al. [4] described a female with a retroperitoneal tumor who was only treated with chemotherapy. She died of disease 24 months after initial presentation. In addition, the case reported by Caruncho et al. [2] did not inform patient's final outcome.

In the present case, patient underwent a radical operation followed by adequate platinum-based chemotherapy, and obtained an excellent clinical outcome. The patient was alive with no evidence of disease for 21 months. Given chemotherapy after recurrence, she survived up to now. The outcome of the patient demonstrated the potential effectiveness and feasibility of this treatment pattern. In the literature, it has been emphasized that patients with positive surgical margins, tumor infiltration of adjacent organs or loco-regional lymph node involvement should be treated with chemotherapy [3,5]. Although more cases are needed, we believe that the therapy modality combining a radical operation with platinum-based chemotherapy may be the top candidate for treatment of PRSA. Meanwhile, we must pay attention to the adverse reactions of chemotherapy. Yonehara et al. [13] have described a woman with primary serous papillary carcinoma (PSPC), after first chemotherapy course, pericardial effusion occurred and a pericardiectomy was performed to prevent cardiac failure.

Conclusion

In conclusion, we report an unusual case of a 58-year-old woman with primary retroperitoneal serous adenocarcinoma, and the postoperative pathology provides powerful evidence for the theory of caelomic metaplasia. Although more cases are needed, we emphasize that radical surgery combined with platinum-based chemotherapy may be the preferred method for treatment of PRSA.

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