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Case Report

Primary peritoneal carcinosarcoma: A report of two cases

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ABSTRACT

Objective: Carcinosarcomas also known as malignant mixed mullerian tumors (MMMTs) contain both carcinomatous and sarcomatous elements. Most MMMTs are arising from female genital tract, including ovaries, uterus and fallopian tubes. Extragenital carcinosarcomas are extremely rare, with an estimation less than 40 cases so far.

Case report: We report two cases of primary peritoneal carcinosarcomas. An 81-year-old woman with pelvic peritoneal carcinosarcoma, heterologous type, was treated with incomplete surgery without further chemotherapy, and died of disease soon. The other one was a 76 year-old woman with abdominal peritoneal carcinosarcoma, homologous type. After optimal debulking surgery and subsequent 6 cycles of combination of paclitaxel and carboplatin chemotherapy, the patient is free of tumor half of year.

Conclusion: Active therapy, including complete cytoreduction surgery and carboplatin-paclitaxel chemotherapy might offer a chance of disease control for these unusual primary peritoneal carcinosarcomas.

Introduction

Carcinosarcoma, a malignant biphasic neoplasm consisting of a carcinomatous component and a malignant non-epithelial component of mesenchymal origin, is also known as malignant mixed mullerian tumor (MMMT). Usually, MMMTs arise from the female genital tract, including the ovaries, uterus and fallopian tubes. Those that arise from extragenital locations, such as the pelvic peritoneum, serosal surface of the colon, retroperitoneum, anterolateral abdominal peritoneum, and omentum, are extremely rare [1]. Moreover, there are less than 40 cases by literature review [3]. The first primary peritoneal carcinosarcoma was defined by Ober and Black [2] in 1955. The primary peritoneal carcinosarcoma is an aggressive tumor. Most patients relapse within one year after completion of initial treatment, with a median overall survival time ranging from 7 days to 73 months [4,5].

We present two cases of primary peritoneal carcinosarcoma. Case 1 originated from the pelvic peritoneum and case 2 originated from anterolateral abdominal peritoneum.

Case presentation

Case 1

A 81-year-old female, G4P4, with body mass index of 25.7 kg/m^2, chronic kidney disease, type II diabetes mellitus, essential hypertension, dementia, and past surgical history of bilateral tubal ligation in 1966 presented at the ER for progressively worsening diffuse abdominal pain with abdominal distension for one week. Physical examination revealed a large firm and non-tender mass at the lower abdomen and pelvic region. Abdominal and pelvic computer tomography (CT) showed a large hypodense pelvic mass, size of 18 cm × 12 cm × 11 cm, displaced bladder and sigmoid colon, and large amount of ascites (Fig. 1A,B). According to the density and location of the mass shown on the CT images, the initial impression made by radiologists was uterine sarcoma. Laboratory data revealed preoperative tumor markers of CA 125 732.1 U/ml and AFP 324.8 ng/ml. The preoperative impression was uterine sarcoma of undetermined origin. Exploratory laparotomy revealed a large neoplasm located above the uterus. The tumor occupied the entire pelvis and extended to the abdominal cavity. The uterus and bilateral adnexa were of normal size. The tumor invaded the serosa of uterus, the sigmoid colon, rectum, pelvic sidewall, and other peritoneal surfaces. Cytoreduction of the pelvic mass was done and intraoperative frozen section was sent for histopathology

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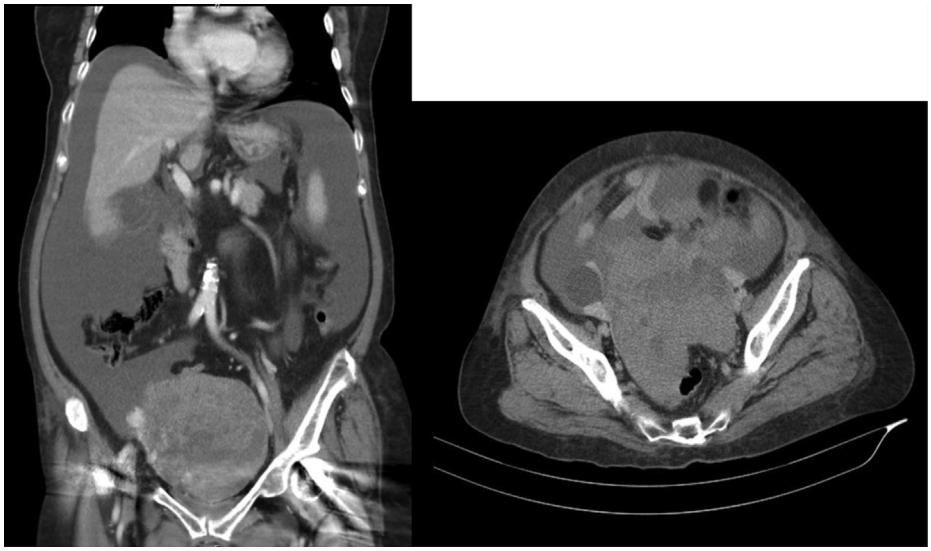

Fig. 1. Computer tomography of abdomen and pelvis showed ascites and a large hypodense pelvic mass, measuring approximately 18 cm × 12 cm × 11 cm, and displaced bladder and sigmoid colon.

diagnosis. Results showed malignant high grade carcinoma Therefore, total hysterectomy, bilateral salpingo-oophorectomy, omentectomy, pelvic and para-aortic lymphadenectomy were performed. Partial colectomy and optimal debulking* surgery were not achieved due to family refusal. The final pathological report was primary peritoneal carcinosarcoma, stage IIIC (T3CN0M0), heterologous type (Fig. 3). Thirty-five days after exploratory laparotomy, follow-up abdominal and pelvic computer tomography showed extensive cancerous peritonitis presented with omental cake, peritoneal seeding and massive ascites (Fig. 2). The patient expired on the 40th day after surgery due to multiple organ failure.

Case 2

A 76-year-old female, G3P3, with body mass index 17.9 kg/m2 and essential hypertension, came to ER due to poor appetite, nausea, and abdominal distension for one week. Abdominal and pelvic computer tomography showed large amount of ascites, and a large hypodense 17 × 15 cm intra-abdominal mass abutting mesentery (Fig. 4). Since the location of the mass did not invade any intra-abdominal organs, the initial impression was to rule out primary peritoneal cancer. Laboratory data showed preoperative CA 125 of 510.5 U/ml. Exploratory laparotomy revealed a multiple cystic tumor up to 17 × 17 cm surrounded by small bowel mesentery with severe adhesion to the terminal ileum. Cytoreduction of the intra-abdominal mass and small bowel mesentery, total hysterectomy, bilateral salpingo-oophorectomy, omentectomy, pelvic and para-aortic lymphadenectomy were performed. Optimal debulking* was achieved. The final pathological report was primary peritoneal carcinosarcoma, stage IIIC (T3CN0M0), homologous type (Fig. 6). Post-operative CA125 was 145.2 U/ml. Combination adjuvant chemotherapy with paclitaxel and carboplatin was administered in 6 cycles. Tumor marker follow-up of CA125 lowered to 17.19 U/ml. Four months after exploratory laparotomy, computed tomography showed no identifiable residual tumor or local recurrency in the peritoneal cavity (Fig. 5). The patient is still alive 6 months after she was first diagnosed.

Discussion

Carcinosarcomas are highly aggressive neoplasms that can be detected in many locations, mainly the female genital tract [6]. Extragenital carcinosarcomas are extremely rare and mostly

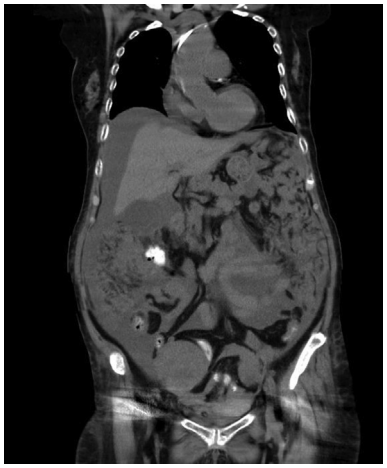

Fig. 2. Computer tomography showed extensive cancerous peritonitis presented with omental cake, peritoneal seeding and massive ascites.

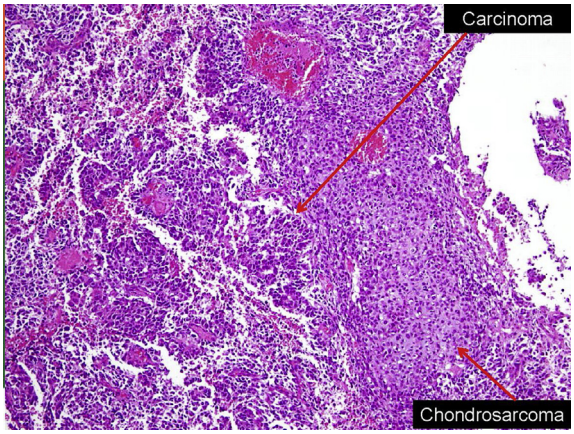

Fig. 3. Pathological picture showed carcinosarcoma, with chondrosarcomatous contents (heterologous type).

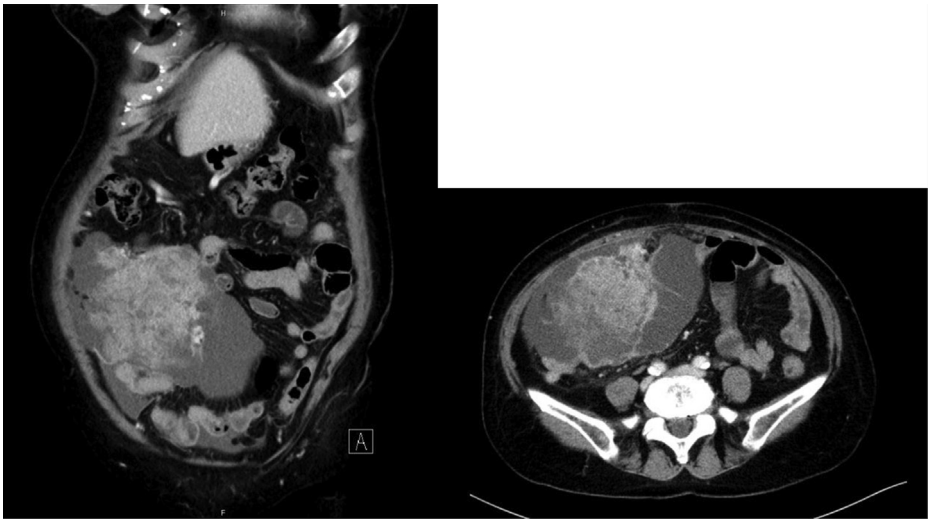

Fig. 4. Computer tomography showed large amount of ascites including a large hypodense 17 × 15 cm intra-abdominal mass abutting mesentery.

develop from the peritoneum, followed by the serosal surface of the colon, retroperitoneum, anterolateral abdominal peritoneum, and omentum [2]. According to previous literature reviews [4], patients with primary peritoneal carcinosarcoma originating from the pelvic peritoneum or uterine serosa has better survival rate than that originating from other peritoneal surfaces.

Carcinosarcoma can be categorized as homologous or heterologous, depending on the histologic characteristics of the sarcomatous elements [7]. Homologous carcinosarcomas have a sarcomatous component of fibrosarcoma, endometrial stromal sarcoma or leiomyosarcoma. Heterologous types include sarcomatous components that are made up of tissues non-native to the uterus such as malignant cartilage or skeletal muscle. According to previous literature reviews [8], mean survival outcome among patients with homologous types was better than those with heterologous types. Likewise, case 2 with homologous carcinosarcoma has a better prognosis than case 1 with heterologous components.

Surgical excision is the main treatment for carcinosarcomas. Optimal debulking surgery, with residual tumor less than 1 cm, should be achieved. In Case 1, cytoreduction of pelvic mass, total hysterectomy, bilateral salpingo-oophorectomy, omentectomy, pelvic and para-aortic lymphadenectomy were performed. Due to family refusal of partial colectomy, optimal debulking surgery could not be achieved. Residual tumors at the sigmoid colon, rectum and pelvic wall relapsed within 1 month.

Due to the rarity of the disease, there is only limited data regarding the management of the disease. The key point of treatment is surgical debulking. However, most cases of carcinosarcoma have widely spread metastasis at the time of presentation, making optimal tumor debulking difficult [9]. Systemic chemotherapy is strongly advised in all cases irrespective of stage at time of diagnosis because of early tumor spread. Platinum in combination with ifosfamide are the preferred agents [10]. Adjuvant radiotherapy after debulking surgery in combination with chemotherapy may be considered for small residual diseases [5]. According to literature reviews, primary carcinosarcomas were found to be linked with synchronous or metachronous gynecologic tumors of mullerian duct origin (ie, ovarian tumors, primary serous carcinoma of the peritoneum, fallopian tube cancer, endometrial cancer), with 37.5% association [11]. Because the tumor behavior is determined by the epithelial component, management of carcinosarcomas should follow that of high-grade serous carcinoma of the ovary [12]. Studies have shown that ovarian carcinosarcoma cases that have received combination chemotherapy of platinum and taxane have longer median survival rate [13,14]. In Case 2, combination adjuvant chemotherapy with paclitaxel and carboplatin was administered in 6 cycles. The CA125 after surgical treatment reduced from 145.2 U/ml to 17.19 U/ml after adjuvant chemotherapy. There is not enough

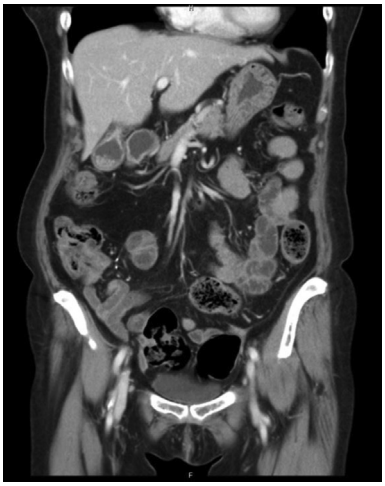

Fig. 5. Computer tomography showed no identified residual tumor or local recurrence in peritoneal cavity.

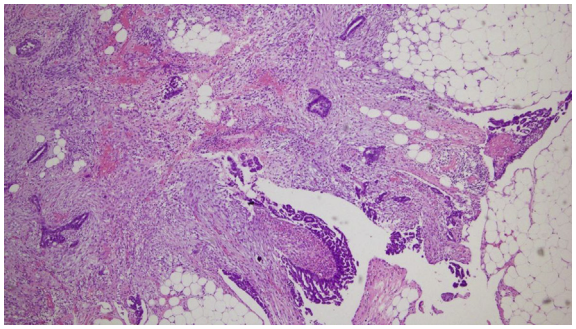

Fig. 6. Pathological picture showed carcinosarcoma (homologous type). *Definition of optimal debulking surgery: residual tumor <1 cm.

data to support the role of radiotherapy usage in extragenital carcinosarcoma [15].

Conclusion

Primary peritoneal carcinosarcomas are extremely rare. There are only limited data regarding their management. Recommendations for the treatment of MMMTs are based on individual cases. According to our cases on two different histological types, optimal debulking surgery and adjuvant chemotherapy could affect survival outcome of these extremely rare diseases.

Conflicts of interest

All authors declare that there is no conflict of interests regarding the publication of this paper.

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