Annals of Clinical Case Reports

Case Report
Published: 26 Mar, 2019

Anaplastic Ovarian Carcinoma Arising From a Background of Mucinous Carcinoma with Fatal Outcome: Case Report and Literature Review

Ji-Young Hong[1], Sujin Park[2], Tae-Joong Kim[1], Sang Yong Song[2], Jeong-Won Lee[1], Byoung-Gie Kim[1], Duk-Soo Bae[1], Chel Hun Choi[1] and Nam Rri Kim[1]*

[1]Department of Obstetrics and Gynecology, Sungkyunkwan University School of Medicine, South Korea

[2]Department of Pathology and Translational Genomics, Sungkyunkwan University School of Medicine, South Korea

Abstract

Ovarian mucinous carcinoma is 2% to 4% of all epithelial ovarian cancers and is known to have good prognosis. It is low stage at diagnosis, but one of the least studied types of ovarian epithelial cancer. Sometimes anaplastic foci are found with ovarian mucinous carcinoma and several types of mural nodules may develop in the wall of mucinous tumors. It is rarely encountered and reported in less than 35 cases with no reports in South Korea. Prognosis of these, if unruptured, is not as poor as initially suggested. We report a case of anaplastic carcinoma arising from mucinous carcinoma with fatal outcome in a 24-year-old female.

Keywords: Mucinous carcinoma; Anaplastic carcinoma; Poor prognosis

OPEN ACCESS

Correspondence:
Chel Hun Choi, Department of Obstetrics and Gynecology, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, South Korea, Tel: 82234103545; Fax: 82234100630;
E-mail: Chelhun.choi@samsung.com

#The first and last 2 authors contributed equally to this paper.

Received Date: *21 Feb 2019*
Accepted Date: *21 Mar 2019*
Published Date: *26 Mar 2019*

Citation:
Kim NR, Park S, Kim T-J, Song SY, Lee J-W, Kim B-G, et al. Anaplastic Ovarian Carcinoma Arising From a Background of Mucinous Carcinoma with Fatal Outcome: Case Report and Literature Review. Ann Clin Case Rep. 2019; 4: 1626.

ISSN: 2474-1655

Introduction

Mucinous carcinoma of the ovary is rare and the least researched type of ovarian epithelial carcinoma. Ovarian mucinous carcinoma is 2% to 4% of epithelial ovarian carcinomas. It usually has a good prognosis and is low stage at presentation [1]. Anaplastic foci are sometimes found within mucinous carcinomas and several types of mural nodules may develop in mucinous tumor walls. Anaplastic carcinoma is very rarely encountered; less than 35 cases have been reported to date. Prognosis is poor, but not as poor as initially suggested if unruptured [2]. We report a case of anaplastic carcinoma arising from mucinous carcinoma with fatal outcome in a 24-year-old female. Although the patient was diagnosed with early (stage Ic) disease, wide spreading of recurrent tumors occurred rapidly after first debulking surgery and three cycles of chemotherapy, consistent with aggressive behavior for this tumor.

Case Presentation

The patient was 24 years old, gravida 0 and para 0, with no underlying disease. She presented with abdominal fullness and dyspepsia. CT scans done at another hospital showed a large abdominal mass of about 30 cm sized arising from the left ovary. In our hospital, pelvis MRI was done and CT scan and pelvis MRI findings showed a large, multiseptated cystic mass. Each locule had varied components including a solid portion, so this finding was potentially indicative of borderline left ovarian epithelial tumor. On presentation to our hospital, the patient felt abdominal fullness with mild tenderness in the right lower quadrant area. Serum tumor markers were within normal limits: CA-125=11.3 U/mL and CA-19-9=11.5 U/mL. No other abnormal findings were consistent with preoperative images. On laparoscopic surgery, a 20 cm × 15 cm left ovarian mass was seen, filled with mucinous content. Pelviscopic left salpingo-oophorectomy, right ovarian fossa biopsy for suspicious peritoneal lesion, and partial omentectomy were done. Frozen diagnosis during surgery was suspicious of malignancy, but cell type was uncertain.

A thorough pathologic exam was requested and staged as a FIGO IC due to surgical spill. Pathologic confirmation was mucinous carcinoma grade I with multiple focal anaplastic carcinoma nodules (Figures 1A-1C). Multiple nodules were found at the cyst wall, which appeared as subtle, multifocal elevations at approximately six foci. Sizes were up to 2.3 cm. No gross surface involvement was found and final diagnosis was anaplastic carcinoma of ovary through immunohistochemistry testing.

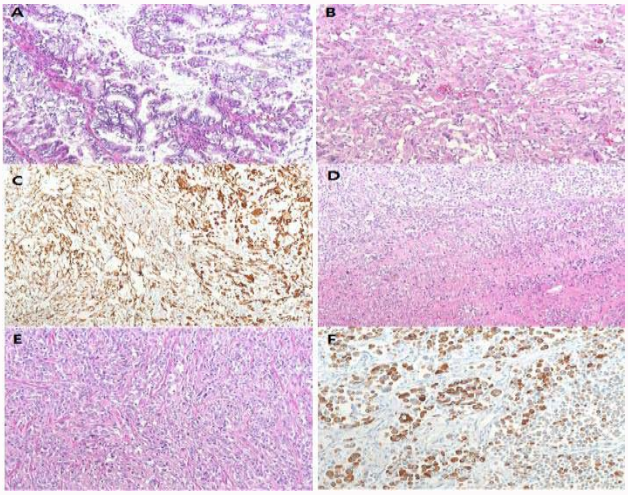

Figure 1: Microscopy of main tumor, anaplastic foci, and metastatic tumor**.** Main tumor and anaplastic foci: (A) Mucinous carcinoma component in main tumor with expansile growth of intestinal type mucinous epithelium and little or no intervening stroma. (B) Nodules were mainly two components: large rhabdoid and pleomorphic cells. The two components were intimately admixed without discrete borders. The pleomorphic-to-rhabdoid ratio was about 7:3 with 18–19 mitotic figures per 10 high power fields. (C) Pleomorphic and rhabdoid components of nodules were positive for cytokeratin staining. Metastatic tumor: (D) Additional necrosis. (E) Spindle cell component with rhabdoid and pleomorphic cell components. (F) Tumor cells were positive for cytokeratin immunostaining. (A), (B), (E), (F) 200x; (C), (D) 100x.

Postoperatively, serum tumor markers were still within normal limits: CA-125=27.9 U/mL and CA-19-9=16.4 U/mL. The patient subsequently underwent three cycles of chemotherapy with paclitaxel and carboplatin. At follow up, serum tumor markers had changed to CA-125=27.7 U/mL and CA-19-9=12.8 U/mL. After three cycles of chemotherapy, the patient complained of mild right low-quadrant discomfort. A follow up CT scan following three cycles of chemotherapy showed multiple seeding implants at Morison's pouch and the subhepatic space, both paracolic gutter and pelvic peritoneum. Debulking surgery was planned because the patient had not received comprehensive staging of debulking at initial surgery. Before the planned surgery, the patient visited the emergency room for abdominal pain, and mechanical ileus was diagnosed. Her pain was aggravated and ileus was aggravated daily.

Due to aggravating symptoms, we did a follow up CT scan 16 days after the last scan. More progressive disease was seen and obstructive ileus was found due to descending colon invasion by the seeding mass. The size of all seeding masses in the perihepatic area increased and new, direct invasion seeding of the lesion to the liver parenchyma was seen (Figure 2). Tachycardia and abdominal pain aggravated with time, but the patient did not have fever. She underwent total abdominal hysterectomy, right salpingo-oophorectomy, and peritonectomy, and appendectomy, total colectomy with ileostomy, splenectomy and seeding mass removal. Intraoperatively, 3L bloody with foul-odor turbid ascites was discovered and a 10 cm left paracolic gutter mass had invaded the descending colon, resulting in spontaneous colon perforation. A 10 cm subhepatic mass had invaded the liver parenchyme and porta hepatis. Ineffaceable seeding nodules were found at the peritoneum, omentum, spleen, subdiaphragmatic space and Morison's pouch. Bulky masses were removed and residual disease measured less than 1 cm.

Pathologic examination showed anaplastic carcinoma and tumor

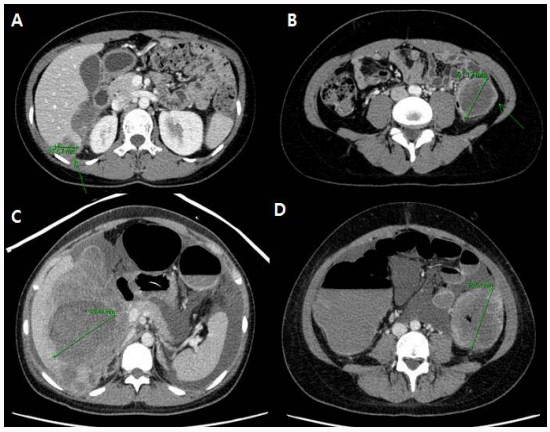

Figure 2: CT at admission for abdominal pain and mechanical ileus and 16 days later. CT at admission: (A,B) Seeding mass at perihepatic area (2.7 cm) and left paracolic gutter (5.2 cm). Follow up CT 16 days later: (C,D) More progressive disease with obstructive ileus due to descending colon invasion by seeding mass. Sizes of all seeding masses in perihepatic area increased (2.7 cm to 9.8 cm) with 7.8 cm new direct invasion seeding lesion in the liver parenchyma.

cells at the right ovary, uterus, peritoneum, perihepatic mass, and appendix, ascending and descending colon, cecum and mesentery (Figures 1D-1F). The specimens showed only anaplastic carcinoma components without mucinous carcinoma component. Cytokeratin AE1/AE3 PAN CK and vimentin were positive in tumor cells, similar to the primary pathologic exam, with PAX8 negative characteristics in tumor cells.

Postoperative ileus developed even though total colectomy and ileostomy were performed. Postoperative ileus suggested the tumor invaded the small bowel and small bowel perforation was assumed. CT showed that increased size of residual masses from debulking surgery at Morison's pouch and the subhepatic and perihepatic spaces. These lesions directly invaded the hepatic parenchyme, porta hepatis and abdominal wall. Operation wound disruption was seen with large amounts of discharge from the wound. Therefore, we performed wound repair in the operation room and found multiple seeding lesions at the wound disruption site. The patient died of respiratory failure and sepsis due to disease progression 13 days after secondary cytoreductive surgery and 4 months after diagnosis.

Discussion

Most mucinous carcinomas are associated with benign mucinous epithelium or mucinous borderline tumors of intestinal type [1]. Mural nodules can be seen in all ovarian mucinous tumors. Mural nodules are rare and classified into Sarcoma-Like Mural Nodules (SLMNs), true sarcomas, and Anaplastic Carcinomas (ACs). SLMNs are well-circumscribed tumors ranging from 0.6 cm to 6 cm. Immunohistochemistry shows focal and weakly positive total cytokeratin [3]. Anaplastic carcinomas often have only foci, unlike SLMNs that usually have one or several nodules and invade microscopically, which is why they demonstrate poor demarcation, and range from 1 cm to 10 cm [3, 4]. Rarely, this can show multiple foci [3]. True sarcoma types are exceptionally rare and have poor prognosis [4,5]. They usually resemble fibrosarcoma or undifferentiated sarcoma and have invasive borders [5]. SLMNs are usually confined to the ovary at surgery; however anaplastic carcinomas spread beyond the pelvis [4].

Anaplastic foci in ovarian mucinous carcinomas often have mixtures of carcinoma components [4]. In our case, a large ovarian cyst was removed at initial operation that consisted of mainly mucinous adenocarcinoma components with focal anaplastic nodules. At second surgery, multiple, extensive metastatic nodules were removed. On pathologic examination, no mucinous adenocarcinoma component was observed and all metastatic nodules showed only anaplastic carcinoma components. Compared to the anaplastic components seen from specimens from the initial surgery, tumor necrosis had increased with more pleomorphic cells at metastatic nodules. The ratio of rhabdoid cells decreased, and the spindle cell component was newly developed. All components were still positive for cytokeratin immunostaining.

Provenza and colleagues histologically categorized foci of anaplastic carcinomas into three groups in a series of 34 cases in 2008: (1) Rhabdoid, with diffuse arrangements of cells with large, bright, eosinophilic cytoplasms, eccentric nuclei, and one or more prominent nucleoli; (2) Sarcomatoids, characterized by spindle cell proliferation with atypical and vesicular nuclei often with a herringbone pattern; and (3) Pleomorphic, exhibiting overlapping features of the first two categories [2]. In our case, nodules were composed of mainly two components: one was large rhabdoid cells and the other was pleomorphic cells. The two components were mostly intimately admixed without discrete borders. Therefore, categorizing by the Provenza method was not appropriate in this case. We expect that other cases in the future will not be grouped adequately. A new method for grouping the foci of anaplastic carcinomas is needed.

When initially reported, mucinous carcinomas with anaplastic carcinomatous components were thought to have poor prognosis. Extraovarian spread is associated with an aggressive course [2,4]. However, some studies suggest that anaplastic foci within mucinous carcinoma tumors confined to the ovary do not change prognosis [6]. In the case series reported by Provenza and colleagues, 10 of 15 patients diagnosed with staged Ia disease were alive and clinically free of disease after a mean follow up of 5 years. Six patients were staged with IC disease and half died of the disease after a median time of 8 months [2]. This finding shows that even when confined to the ovary, ruptured or lack of rupture results in substantially different prognoses.

References

1. Gurung A, Hung T, Morin J, Gilks CB. Molecular abnormalities in ovarian carcinoma: clinical, morphological and therapeutic correlates. Histopathology. 2013;62(1):59-70.
2. Provenza C, Young RH, Prat J. Anaplastic carcinoma in mucinous ovarian tumors: a clinicopathologic study of 34 cases emphasizing the crucial impact of stage on prognosis, their histologic spectrum, and overlap with sarcomalike mural nodules. Am J Surg Pathol. 2008;32(3):383-9.
3. Mhawech-Fauceglia P, Ramzan A, Walia S, Pham HQ, Yessaian A. Microfocus of anaplastic carcinoma arising in mural nodule of ovarian mucinous borderline tumor with very rapid and fatal outcome. Int J Gynecol Pathol. 2016;35(4):348-51.
4. Vella J, Cracchiolo B, Heller DS. Anaplastic carcinoma arising in an ovarian mucinous cystadenocarcinoma in a 17-year-old female. J Pediatr Adolesc Gynecol. 2006;19(1):39-43.
5. Philip Clement Robert Young. Atlas of gynecologic surgical pathology. 3rd ed. Elsevier. 2013.
6. Hillesheim PB, Farghaly H. Anaplastic spindle cell carcinoma, arising in a background of an ovarian mucinous cystic tumor: a case report with clinical follow up, review of the literature. Int J Clin Exp Pathol. 2010;3(8):808-11.