

Ovarian mucinous cystadenocarcinoma of low malignant potential associated with a mature cystic teratoma

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ABSTRACT

The development of mucinous cystadenocarcinoma of low malignant potential in a mature cystic teratoma is rare. We report a 36-year-old single female presented with abdominal distension and was found to have a huge pelvic/abdominal mass. Ultrasound revealed a huge cystic ovarian mass with no ascites. Laparotomy and left oophorectomy was performed to the mass. Histology revealed mucinous cystadenocarcinoma of low malignant potential in a mature cystic teratoma.

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Ovarian tumors are classified, according to World Health Organization in 1993, into surface epithelial-stromal tumors (65-70% of overall ovarian tumors frequency), sex cord-stromal tumors (5-10%), germ cell tumors (15-20%), malignant not otherwise specified and metastatic non-ovarian cancer (5%).^{1,2} The mucinous tumor is one category of surface epithelial-stromal tumors of the ovary and it represents approximately 25% of all ovarian neoplasms. It occurs principally in middle adult life. Eighty percent are benign or borderline and approximately 15% are malignant.² Approximately 30-50% of ovarian borderline tumors and 6-10% of ovarian carcinoma are of mucinous type.³

Mature cystic teratoma is the most common ovarian germ cell neoplasm.² The association of ovarian mucinous neoplasm in general with cystic teratoma is infrequent. We report a case presented with a large ovarian cystic lesion composed of 2

components, mucinous cystadenocarcinoma of low malignant potential and mature cystic teratoma.

Case Report. A 36-year-old single lady presented with the history of progressive abdominal distension associated with mild dull abdominal pain. It was not associated with nausea, vomiting, change of bowel habit or weight loss. The menarche was at age 16 years, since that she is having irregular cycles every 2 months for 3 days, not associated with heavy bleeding. Review of other systems was unremarkable. She had no previous medical or surgical illness. She did not receive any hormonal therapy before and not on any medication. There was no family history of malignancy. On physical examination, she was generally well. Abdominal examination revealed a huge non-tender pelvic-abdominal mass up to xiphisternum, which felt firm and regular in contour. Ascites was not detected. Laboratory tests

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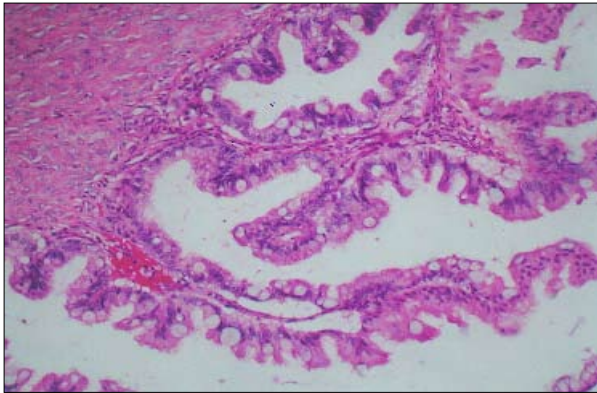


Figure 1 - Borderline mucinous cystic neoplasm. It is lined by mucin secreting epithelium with goblet cells.

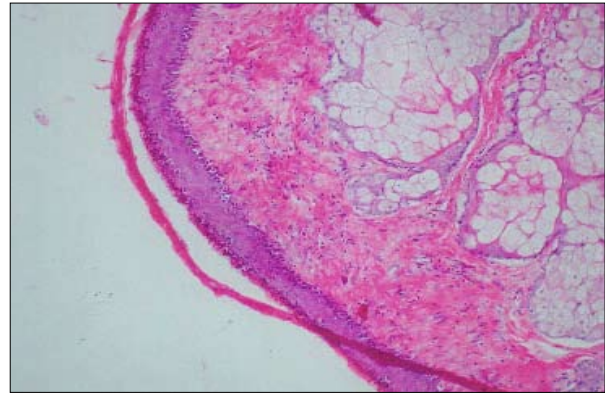


Figure 2 - Ectodermal component of the mature teratoma. Stratified squamous epithelium with underlying sebaceous glands and skin adnexa.

including complete blood count and liver function test were within normal limits. Tumor markers which included; human chorionic gonadotropin, lactate dehydrogenase, alpha-fetoprotein, and cancer antigen-125 (CA-125) were within normal limits. Ultrasound revealed anteverted uterus measuring $6.5 \times 2.5 \times 3.8$ cm, thin endometrium measuring 0.27 cm, and huge hypoechoic homogenous pelvic abdominal mass measuring $16 \times 15 \times 9.3$ cm. There was also complex mass noted in right adnexa measuring $5.6 \times 6.7 \times 6.9$ cm. There was no ascites. She was prepared for laparotomy through a Pfannenstiel incision. There was a huge pelvic-abdominal mass arising from left adnexa reaching the xiphisternum. There was no ascites or any signs of metastasis on the surface. Attach to it was a small cystic mass with a smooth surface. The mass was arising from left adnexa. The right ovary, tube and uterus looked normal. It was difficult to deliver the mass through the incision so purest suture was carried out, a suction was inserted through the cyst, and large amount of sebaceous material and hair was removed (approximately 1000 cc). The ovarian cyst was then delivered out and proceeded for oophorectomy. Abdominal irrigation was carried out. The abdominal wall was closed. The specimen was sent for histopathology.

Macroscopically, this ovarian mass was formed of 2 linked cystic balls. The outer surface of both was smooth, glistening, and gray-whitish in color. The largest cystic ball measured $17 \times 15 \times 8$ cm. On serial sectioning, it was unilocular and filled with a thick mucoid fluid that was focally adherence to the inner wall. The cavity part was showing few papillary projections which were covered by adherent thick mucoid material. The smaller cystic ball measured $7 \times 7 \times 5$ cm. On serial sectioning, it was multilocular and filled with sebaceous, thick, yellowish fluid admixed with hair.

On microscopic examination, the large cystic part was showing several papillary projections, lined by mucin-secreting epithelium with goblet cells (**Figure 1**). These papillary projections were seen anastomosing with each other forming a small cribriform architecture in some areas, while in other areas showing villiform-pattern appearance resembling those of bowel mucosa. No marked cellular atypia or expansile cribriform areas were seen. Extensive sampling failed to show evidence of stromal invasion. The appearance of this cystic neoplasm was in keeping with the mucinous cystic tumor of borderline malignancy. The small cystic part was lined by stratified squamous epithelium with underlying sebaceous glands and skin adnexa (**Figure 2**). Foci of neural elements and small glands lined by intestinal type epithelium were seen. The sebaceous materials as well as the hair shafts were seen inducing foreign body reactive changes with lipogranulomatous formation. This part of the cyst was in keeping with dermoid cyst. A panel of antibodies (immunohistochemistry) were utilized on sections from mucinous neoplasm including: cytokeratin 7, cytokeratin 20, and carcinoembryonic antigen immunostain that showed diffuse strong stain for all the markers.

The patient was discharged after 3 days without uneventful postoperative period. She was referred to the oncology clinic after the final histopathology report and as she was unstaged intraoperative and based on the operative noted and clinic histopathology, she was staged as I-c borderline mucinous tumor which needs close follow up with no chemotherapy at this point.

Discussion. This large ovarian cystic lesion is unique as it is composed of 2 components. One is both grossly as well as microscopically showing classic features of mature cystic teratoma; however,

the larger part showed microscopic features best classified as borderline mucinous cystic neoplasm. The association of ovarian mucinous neoplasm in general with cystic teratoma is approximately 5% of cases,¹ but on literature search, few cases of mucinous cystadenocarcinoma of low malignant potential (as one category of ovarian mucinous neoplasm) associated with mature cystic teratoma in particular have been reported. The pathogenic relationship between these 2 lesions remains unanswered. Although the origin of ovarian mucinous neoplasm is still controversial, evidences point to diverse origins.¹ In most, it is believed that the mucinous part is in fact representing teratomatous mucinous neoplasm; namely, arise from the endodermal component of the teratoma rather than confession tumor. This belief has come from many findings like demonstration of intestinal-type lining in many of these tumors (namely, it will be positive immunohistochemically for cytokeratin 18 and 20, carcinoembryonic antigen, and the peanut agglutinin and negative for α -fetoprotein, cytokeratin 7, CA-125, CA19-9, and human milk fat globulin 1),^{1,4} microscopic appearance resembling for gastrointestinal epithelium and from many reported cases of mucinous tumors arising in ovarian mature cystic teratoma associated with pseudomyxoma peritonei while the appendix is microscopically normal.^{1,3,5,6} The latter refers to 2 tumors from different cell of origin; namely, one as a germ cell tumor and the other is a surface epithelial neoplasm, as the latter will be positive for cytokeratin 7 and negative for cytokeratin 20.⁶

In our case, we suggest more that mucinous neoplasm represents teratomatous mucinous neoplasm; namely, arising from teratoma rather than being separate surface epithelial neoplasm colliding

with teratoma. This is supported by its morphological appearances that resemble gastrointestinal epithelium (the present of goblet cells and villiform-pattern of the epithelium) and immunohistochemical staining results (positive for cytokeratin 20 and carcinoembryonic antigen), although cytokeratin 7 is also positive which cannot be explained.

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