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Case Report

Giant mucinous cystadenocarcinoma in a young unmarried female: a case report

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ABSTRACT

Ovarian epithelial tumours in young females are usually benign, while malignant tumours are extremely rare in this age group. Most of the tumours in young females are usually germ cell-derived. Very few cases of giant mucinous cystadenocarcinoma in young females have been reported till now. Here we present a case of a 23-year-old unmarried female with complaints of diffuse abdominal pain for 4 months and abdominal distension for 3 months. Her contrast enhanced computed tomography (CECT) abdomen was done with an ill-defined large abdomino-pelvic multiloculated solid-cystic mass lesion with heterogeneously enhanced solid component measuring $\sim 21 \times 17.4 \times 10.1$ cm, likely arising from the right adnexa. Her tumour markers were normal. The staging laparotomy was done. Intraoperatively, specimen was sent for frozen section, and the report was suggestive of mucinous cystadenocarcinoma of the ovary. This case report is aimed at showing that large ovarian masses in young females should be evaluated and that timely management should be done to improve the outcome and prognosis.

Keywords: Epithelial tumours, Mucinous cystadenocarcinoma, Tumour markers, Staging laparotomy

INTRODUCTION

Mucinous cystadenomas can arise from teratomas and Wolffian epithelium, or from inclusions and invaginations of the ovarian celomic epithelium and persistence of Müllerian cells.¹ Mucinous cystadenocarcinomas affect people between the ages of 45 and 65 and are only one-third as common as serous cystadenocarcinomas.²

Specifically, mucinous adenocarcinoma has been thought more likely to have a gastrointestinal origin.³ Primary mucinous cystadenocarcinomas are extremely rare, making up about 3-4% of all ovarian cancer cases. The majority of mucinous cystadenocarcinomas are metastatic tumours.⁴

Here we present a case of a giant mucinous cystadenocarcinoma in a 23 years old unmarried female. This case has been reported in line with the SCARE criteria.⁵

CASE REPORT

A 23 years old, unmarried female presented to AIIMS, Rishikesh, India, gynaecology outpatient department with the complaints of pain in abdomen for 4 months, diffuse all over abdomen, on and off, aggravates while micturition. She also complained of abdominal distension for 3 months. She had weight loss (4 kg in 2 months). She had no complaints of loss of appetite/fever/shortness of breath/any menstrual irregularities/PV discharge. Patient had visited private hospital, Moradabad with the same complaints where routine Investigations were done and ultrasonography (USG) abdomen was done s/o right sided moderate hydronephrosis seems to be due to pressure effect. Uterus normal shape and size $7.2 \times 3.8 \times 2.7$ cm, large abdominopelvic cystic mass lesion with septations simulating ovarian cyst? Most probably left side and USG-guided biopsy from right adnexa was s/o well differentiated adenocarcinoma of ovarian origin possibly

mucinous cyst adenocarcinoma. She was then referred to AIIMS Rishikesh for further management.

Her contrast enhanced computed tomography (CECT) (thorax+abdomen+pelvis) was done s/o ill-defined large abdomino-pelvic multiloculated solid-cystic mass lesion with heterogeneously enhancing solid component measuring ~ 21×17.4×10.1 cm is noted likely arising from right adnexa. right ovary is not visualised separately with grossly dilated right ovarian vein. Anteriorly, it is abutting the anterior abdominal wall with indistinct fat planes. Laterally, it is abutting and displacing the small bowel loops with ill-defined fat planes. Posteriorly, it is abutting the bilateral psoas, bilateral common iliac vessels, descending aorta with focal loss of fat planes. It is abutting bilateral ureters with right sided mild hydronephrosis. Superiorly, there is ill-defined fat planes with the transverse colon. Inferiorly, there is loss of fat planes with the uterus. Left inferolaterally abutting the sigmoid colon with ill-defined fat planes. Left ovary is separately visualised. Diffuse omental stranding with nodularity is seen. Gross ascites is seen. Her tumour markers were normal (CA.124 U/ml: 10.30, CA.19.9: 14.90 U/ml, CEA: 1.25 U/ml, AFP: <0.9 ng/ml).

On the initial physical examination, the vital signs were within normal limits. On per abdominal examination, a ~24-26 weeks size, cystic abdominopelvic mass was felt with smooth surface, regular margin, with lower border could not be reached, mobile from side to side and overlying skin normal. Per speculum and per vaginal examination was not done as the patient was unmarried. On per rectal examination, rectal mucosa and bilateral parametria were free and also no nodularity was felt in the cul de sac.

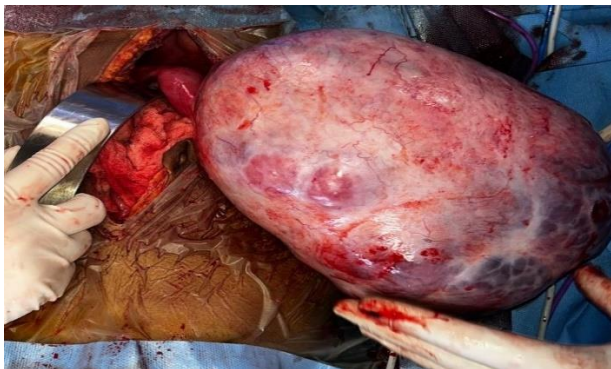


Figure 1: Intraoperative picture of giant ovarian mass.

Patient underwent staging laparotomy. Around 1200 cc of ascitic fluid was aspirated and sent for cytological examination for malignant cells. A 20×15 cm right adnexal mass noted with intact capsule intact and no surface deposits (Figures 1 and 2) but omentum adhered to upper portion of mass.

Right salpingo-oophorectomy and sent for frozen section. Subsequently the patient underwent pelvic and para-aortic lymph node sampling, total omentectomy and appendectomy, conserving her fertility. All the samples were sent for final histopathological examination and the report came to be mucinous cystadenocarcinoma (Figures 3-6).



Figure 2: Cut-section of the ovarian mass.

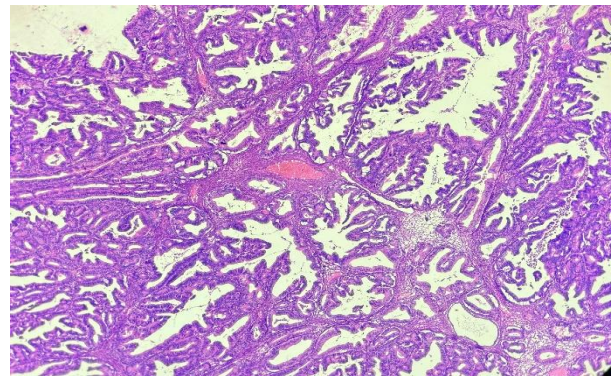


Figure 3: H&E, x100 section showing complex papillary arrangement of glands lined by columnar mucinous cells showing pseudo stratification. The glands are arranged back-to-back with minimal stroma in between.

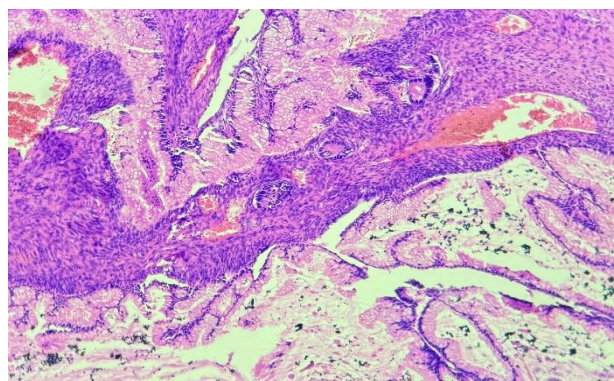


Figure 4: H&E, x200 section showing invasion of neoplastic glands in the stroma (center).

The patient was discharged in stable condition with advice to follow up in medical oncology department for adjuvant chemotherapy.

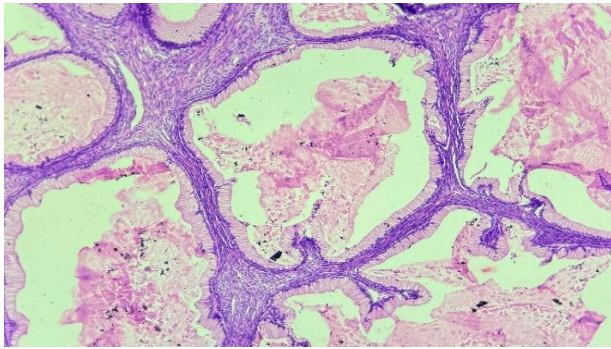


Figure 5: H&E, x200 section showing neoplastic glands lined by columnar cells with basal nuclei and apical cytoplasmic mucin. Intraluminal mucin secretion in the glands is also noted.

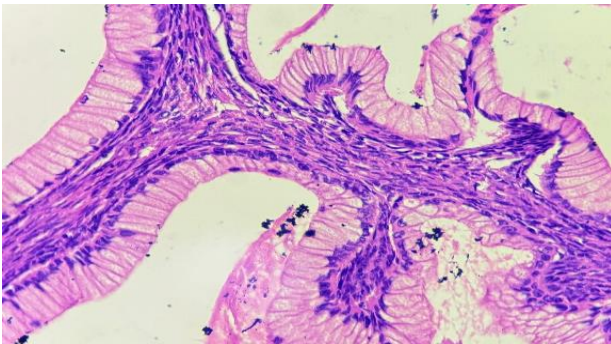


Figure 6: H&E, x400 section showing high power view of the mucinous lining.

DISCUSSION

Ovarian cancer has several pathologic subgroups, the most common of which is epithelial ovarian cancer, accounting for about 80% of cases.³ Adults often develop ovarian epithelial tumours, with a peak incidence of development occurring in those over 40.⁶ Ovarian cystadenocarcinoma is a tumour derived from ovarian epithelial cells and accounts for approximately 60% of ovarian malignant tumours.⁷ Mucinous cystadenocarcinomas may arise de novo or transformation from a benign mucinous cystadenoma which is estimated around 12% of ovarian cystadenocarcinomas.² They are usually unilateral ovarian masses with a large volume, smooth surface, multi-locular cysts/ solid regions, and cystic components; they may be accompanied by bleeding and necrosis.⁴ The majority of patients with these tumours are females in their 20s and 40s. However, cases have also been reported in adolescents, premenarchal girls, and postmenopausal patients.⁸ Giant mucinous cystadenocarcinomas are very rare.⁹

Here we have reported a case of giant mucinous cystadenocarcinoma in a 23 years old unmarried female.

Similar to this, Zhu et al reported a case of giant mucinous cystadenocarcinoma (40×35×34 cm) in a 21 years old female with complaints of abdominal distension for 2 months. Her CEA was highly raised- 199,28806.09 U/ml. Because of the patient's young age and the family's stated wish to maintain fertility, it was treated during the surgical intervention by right adnexectomy, larger omentectomy, and peritoneal biopsy.⁴ Another case of giant mucinous cystadenocarcinoma (6.7×18.9×29.3 cm) was reported by Li et al but in a 14 years old girl with complaints of bloody vaginal discharge. Her tumor markers were within normal limits and was managed by left salpingo-oophorectomy followed by frozen tissue sectioning and subsequently, the patient underwent appendectomy and omentectomy.⁶ Similarly, another case of giant mucinous cystadenocarcinoma (20×17 cm) was reported earlier Gupta et al in a 14 years girl with metastasis to omentum. It was managed by left sided salpingo-oophorectomy and frozen section and subsequently infracolic omentectomy.¹⁰

Reddy et al reported a case of giant mucinous cystadenocarcinoma (20×18×16 cm) with cavitary metastasis to lungs in a 55 years old multiparous postmenopausal female with the complaints of abdominal pain and progressively increasing abdominal distension for 3 months with occasional episodes of hemoptysis. Her CA 125 levels were found to be elevated at 970 IU/ml, while carcinoembryonic antigen and alpha-fetoprotein were normal. The patient underwent exploratory laparotomy with debulking of the left ovarian mass, total abdominal hysterectomy, and bilateral salpingo-oophorectomy. Histopathology examination of the resected specimen confirmed the diagnosis of ovarian mucinous cystadenocarcinoma. Six cycles of adjuvant chemotherapy have been administered 3 weeks apart with cisplatin plus paclitaxel. Thoracoscopic upper lobectomy of the left lung was performed 3 weeks after the last chemotherapy and histopathological analysis of frozen sections from the resected specimen confirmed mucinous cystadenocarcinoma.¹¹ Another case of postmenopausal female with giant mucinous cystadenocarcinoma (36×34×25 cm) was reported by Sengupta et al. Her CA-125 was 66.57 U/ml. The patient underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy, pelvic lymphadenectomy, partial omentectomy and received six cycles of adjuvant chemotherapy.⁸

CONCLUSION

Mucinous cystadenocarcinomas can even present as giant abdominopelvic masses in premenarchal girls, adolescents or old aged women. Fertility sparing surgery should be considered in young females. Adjuvant chemotherapy should be considered if necessary and proper follow up of the patients should be done.

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Ethical approval: Not required

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