**Bilateral mucinous ovarian carcinoma in a young female with tumor weighing 41.1 kg**

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**ABSTRACT**

Mucinous ovarian carcinoma is a less-prevalent subtype of epithelial tumors. We present a case of giant mucinous ovarian carcinoma weighing 41.1 kg in a 24-year-old Indian girl. Benign mucinous tumors have the potential to reach an enormous size but such a huge malignant mucinous tumor is very rare in literature and its occurrence in a young female is even rarer.

**Keywords:** 41.1 Kg, bilateral mucinous ovarian carcinoma, huge ovarian tumor, young Indian girl

**Introduction**

Epithelial ovarian carcinoma (EOC) is the most common histopathological variant of ovarian malignancy. It predominantly occurs in postmenopausal women; merely 10%–15% of EOCs are reported in premenopausal women.¹ Furthermore, <1% of them occur in women younger than 30 years.²

Mucinous ovarian carcinoma is a comparatively less-prevalent subtype of EOCs (3%–5%) and only 10% of them are bilateral.³ Mucinous ovarian tumors are slow-growing tumors and have the potential to grow enormously. Most of the reported cases of huge ovarian mass are benign, mucinous cystadenoma [Table 1].³⁴ It is quite uncommon for a malignant ovarian tumor to be presented as a huge abdominal mass; primary care physicians should be vigilant not to miss an ovarian malignancy as one of the possible causes and should not delay referral to the gynecologist or oncologist.

We present a case of giant mucinous ovarian carcinoma weighing 41.1 kg (90.61 pounds) in a young Indian girl.

**Case Presentation**

A 24-year-old unmarried girl presented with abdominal distension and dull aching pain abdomen for 6 years with difficulty in walking, standing, and breathing for 1 year. She was cachexic, anemic, and hypoproteinemic, weighing 78.8 kg on presentation. The abdomen was grossly distended from epigastrium to hypogastrium [Figure 1a]. A huge cystic to firm abdominopelvic mass with ill-defined borders was palpated. CECT abdomen and pelvis [Figure 1b] showed a large multiloculated cystic lesion (51 cm × 40 cm × 30 cm) occupying the whole abdominopelvic cavity with right gross hydroureteronephrosis. Tumor marker evaluation revealed raised CA-125 (160 U/L), CA19-9 (9314 U/mL), CEA (253 ng/mL), LDH (456 U/I), normal AFP, and Beta-HCG. Upper gastrointestinal (GI)
endoscopy and colonoscopy were normal, done in view of raised CA19-9, and to rule out primary GI malignancy.

A multidisciplinary team was formed and planned for staging laparotomy with the consent of intraoperative and postoperative morbidity and mortality. Preoperative right-sided percutaneous nephrostomy (PCN) was done in view of gross hydroureretonephrosis. Anemia and hypoproteinemia were corrected.

On laparotomy, a 60 cm × 50 cm × 40 cm multiloculated mass adhered to the abdominal wall was noted with large omental feeding vessels. Mass was gradually decompressed as advised by the anesthetist to avoid sudden hemodynamic instability. During aspiration, cardiopulmonary parameters did not change. Precaution to avoid tumor spillage was taken. Approximately 36 liters of thick straw-colored mucinous fluid was drained. After adhesiolysis, mass was seen arising from the left ovary [Figure 1c]. Left adnexectomy was performed. The specimen weighed 5.1 kg, making the total weight of ovarian mass 41.1 kg. A 5 cm × 4 cm simple cyst in the right ovary and multiple tumor deposits (~1 cm) in Pouch of Douglas were also noted. The uterus was normal in size with no tumor deposits. Urinary bladder, omentum, undersurface of liver, diaphragm, spleen, large and small bowel were free of tumor. Bilateral pelvic and paraaortic lymph nodes were not palpable. The parietal and pelvic peritoneum was grossly normal. Intraoperative imprint cytology of left ovarian mass was suggestive of mucinous cystadenocarcinoma. Therefore, complete cytoreductive surgery including total abdominal hysterectomy, right salpingo-oophorotomy, total omentectomy, bilateral pelvic lymph node dissection, appendectomy, peritoneectomy and removal of POD tumor deposits was performed. Estimated blood loss was 4.5 liters. The surgery lasted for 6 hours and patient was transferred to Intensive Care Unit (ICU) for cardiopulmonary support. She received massive blood transfusion in the perioperative period and had a prolonged ICU stay. Approximately 60% of her bodyweight was removed in the form of tumor, and she took almost one month to recover. In immediate postoperative period, the patient was kept on ventilatory support and inotropes as she was not able to maintain intravascular volume. Broad-spectrum antibiotics were given to cover both aerobic and anaerobic organisms. On postoperative day (POD) 2, she developed sudden hypotension and bradycardia followed by asystole; prompt cardiopulmonary resuscitation was given and revived successfully. She had an episode of paroxysmal supra-ventricular tachycardia on POD 4, managed with amiodarone. She developed metabolic derangements i.e., hypokalemia, hypocalcemia, and hypomagnesemia; 2D echocardiography showed global left ventricular hypokinesia, LVEF-20%, mild MR, and mild TR. Electrolyte correction was given accordingly. Injection albumin was given daily for 10 days to correct hypoalbuminemia. On POD 7, patient was hemodynamically better and extubated and inotropic support was gradually weaned off. But next day only, she was not maintaining saturation on noninvasive ventilation and was electively intubated. Followed by this, she developed subcutaneous emphysema over her face and neck, airway injury was suspected; urgent CT thorax suggested left gross pneumothorax and gross pneumomediastinum [Figure 2a] for which emergency intercostal drain (ICD) insertion and CT guided pigtail insertion in pneumomediastinum was performed [Figure 2b]. Patient was restarted on inotropes. Tracheostomy was performed on POD 10 in view of prolonged intubation and difficulty in weaning.

**Table 1: Cases reported in the Literature of Huge Ovarian Tumor**

| Author, Year | Age (years) | Pre/ postmenopausal | Tumor Weight (kg) | HPE    | HPE type of ovarian tumor | Outcome |
|-------------|-------------|---------------------|------------------|--------|-------------------------|---------|
| Akhras et al. (2019) | 72 | Postmenopausal | 27 | Benign | Mucinous cystadenoma | Fair |
| Gwanzura et al. (2019) | 48 | Postmenopausal | Occupying whole abdominal cavity | Benign | Mucinous cystadenoma | Died |
| de Lima et al. (2014) | 57 | Postmenopausal | 40 | Malignant | Mucinous cystadenocarcinoma | Fair |
| Madhu YC, et al. (2013) | 55 | Postmenopausal | 56.95 | Benign | Mucinous cystadenoma | Fair |
| Fobe D, et al. (2010) | 24 | Premenopausal | 59 | Benign | Mucosal-serosal cystadenoma with borderline potential | Fair |
| Hackerth et al. (2008) | 60 | Postmenopausal | 60 | Benign | Mucinous cystadenoma | Fair |
| Yanazume Y, et al. (2007) | 30 | Premenopausal | 100 | Malignant | Mucinous cystadenocarcinoma | Died |
| Present Case | 24 | Premenopausal | 41.1 | Malignant | Mucinous cystadenocarcinoma | Fair |

HPE—Histopathology
Khoiwal, et al.: A giant malignant ovarian tumor in a young girl

Patient started improving gradually, inotropes weaned off on POD 15, ICD and pigtail were removed on POD 19. Tracheostomy tube was removed on POD 22. Repeat 2D echocardiography was suggestive of normal left ventricular function. PCN was removed as hydroureteronephrosis was resolved in review scan. Patient developed grade III/IV bed sores due to prolonged immobilisation which were recovered by daily dressing, improved diet and ambulation. Her body weight on the day of discharge was 31.3 kg.

Final histopathology [Figure 1d] was suggestive of bilateral mucinous carcinoma of ovary with left parietal peritoneum and pouch of Douglas involvement - FIGO stage IIIA (immunohistochemistry; ER and PR - negative, p53 - diffuse nuclear positivity, CDX2 - nuclear positivity, CK20 and CK7 - positive). She is currently on platinum-based adjuvant chemotherapy and received 9 weekly cycles uneventfully.

Institutional Ethical Board approval (AIIMS/IEC/20/643) and consent from the patient was obtained for publication.

Discussion

Our case was unique due to the occurrence of bilateral mucinous ovarian carcinoma in a young female with a tumor weighing 41.1 kg. A similar case of giant mucinous ovarian malignancy was reported in a 30 years old woman by Yanazume (2007) but unfortunately, patient could not withstand surgery and died 12 hours postoperatively due to massive blood loss and DIC.[9]

Giant ovarian tumors alter the patient’s anatomy, physiology and psychology. Pressure symptoms, elevation of diaphragm, compromised lung capacity, impaired venous return and decreased cardiac output are caused by such huge intra-abdominal masses. Furthermore, removal of these tumors are associated with innumerable complications. Cardiovascular problems are the most common followed by respiratory complications and splanchnic shock. In our case too, patient developed sudden hypotension followed by cardiac arrest on POD 2, fortunately she could revived successfully. Monitoring in ICU, early recognition of complications and immediate management are pivotal for optimum outcome.

Ideally in such cases, an attempt should be made to remove the mass intact to avoid dissemination of tumor but we decompressed the tumor gradually to avoid sudden hemodynamic instability. Guraslan H, et al.[10] contemplated controlled fluid aspiration before or during surgery to prevent risk associated with sudden intraabdominal pressure changes.

In early stage, prognosis remains good but advanced staged tumors have a poor prognosis as mucinous tumors are relatively chemoresistant. Hence, cytoreduction should be optimum. Fertility sparing surgery should be considered in young patients in stage I but not beyond stage Ic3.[11] It was not an option in our case as intraoperative findings were suggestive of stage IIb and moreover, patient and her mother were keen for complete surgery in one go and not willing for a second surgery.

To the best of our knowledge, this is the largest malignant ovarian tumor being reported from India and the first of its kind in the world being presented at such a young age with a 41.1 kg ovarian malignancy and survived after the surgery. Preoperative optimisation, anticipation of complications and monitoring in intensive care unit are crucial for a fair outcome.

Key points

- Rare histopathological type: Mucinous ovarian carcinoma is a less prevalent subtype of Epithelial ovarian carcinoma (3%–5%).
- Bilaterality: It was a case of bilateral mucinous ovarian carcinoma, only 10% of mucinous ovarian malignancies are bilateral.
- Age of presentation: Most of the mucinous ovarian cancers present in postmenopausal women but in our case it was presented in a young 24 year old female.
- Size: Most of the large ovarian tumors reported in literature are benign in nature (mucinous cystadenoma) but it was malignant in our case (mucinous cystadenocarcinoma).
- Prognosis: This is the largest malignant ovarian tumor being reported from India and the first of its kind in the world being presented at such a young age with a 41.1 kg ovarian malignancy and survived after the surgery.

Ethical committee approval

The study has been approved by the Institutional Ethical Board - AIIMS/IEC/20/643.

Declaration of patient consent

Consent for publication was obtained from the patient.

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Conflicts of interest

There are no conflicts of interest.
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