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Case Report

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### Ovarian Serous cystadenocarcinoma in a young woman: a rare occurrence

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#### ABSTRACT

Serous cystadenocarcinoma (SC) is the most common ovarian carcinoma usually affecting elderly females. Exact incidence of SC in females younger than 25 years is not known due to rarity and little available literature. We report a rare occurrence of ovarian serous cystadenocarcinoma in a young nulliparous lady of 22 years age emphasizing the fact that though unusual, possibility of SC should be kept in mind in younger age group owing to its low grade morphology, early stage at the time of diagnosis and excellent prognosis.

**Keywords:** Serous cystadenocarcinoma, Surface epithelial tumor, Ovarian tumor.

#### INTRODUCTION

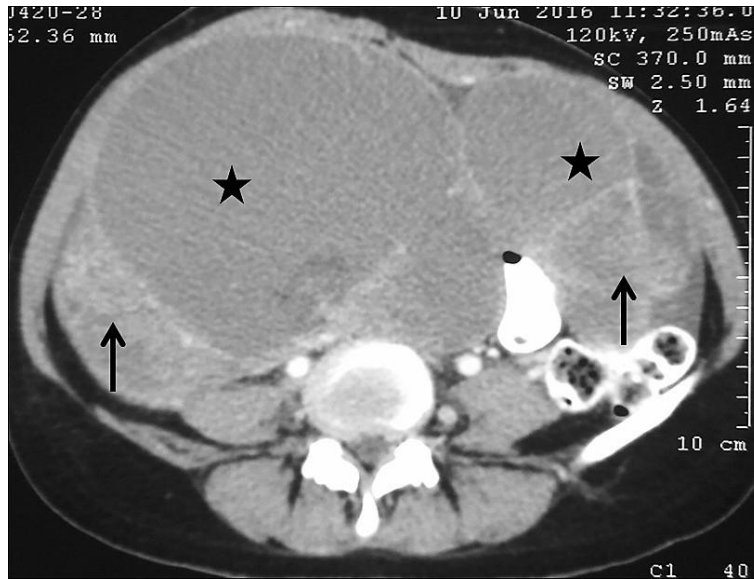
Serous cystadenocarcinoma (SC) is the most common type of ovarian surface epithelial tumor accounting for approximately 50% of all cases. [1] It most commonly occurs in 6<sup>th</sup>-7<sup>th</sup> decade of life, with a reported mean age of 57-63 years. It rarely affects children and adolescents with <1% incidence below 20 years of age. [1, 2] Little literature is available quoting the exact incidence in women below 30 years. Till date, around 35 cases of ovarian serous epithelial carcinomas have been reported below 20 years of age. Most of these were diagnosed at an early stage. [1] We report a case of a 22 years lady with an ovarian mass masquerading as malignant germ cell tumour which turned out as low grade serous cystadenocarcinoma on histopathology.

#### CASE REPORT

A 22 year old nulliparous lady, married for three years, presented to gynaecology outpatient department with chief complaints of intermittent pain lower abdomen for the past 6 months. The pain was dull aching, non-radiating and was not associated with menstruation or bladder/bowel symptoms. Patient also complained of abdominal distension which was gradually increasing in size for the past 6 months. It was associated with nausea and vomiting. There was no history of weight loss, loss of appetite or menstrual complaints. Per abdomen examination revealed, a well defined, mobile, firm mass measuring 20x10cm arising from the pelvis and extending up to the right hypochondrium. Per vaginum and per speculum examination revealed normal cervix and vagina

with no evidence of discharge or bleeding. Routine investigations including CBC, LFT and KFT were within normal limits. CA 125 was markedly raised to 2606 u/ml, CEA was 1.2 U/ml, CA 19.9 was 1.8U/ml and serum LDH was 297 units/l. Ultrasound abdomen showed a large cystic abdomino-pelvic mass, which measured 25x18x10 cm. The mass was crossing the midline, had well defined margin, few thick septae and multiple

internal papillary projections. Ovaries were not seen separately. Contrast-enhanced computed tomography (CECT) abdomen showed a large, abdominopelvic multiloculated and multiseptate mass measuring 26x18x10.9cm with few enhancing solid components. The mass seemed to be arising from bilateral adnexal regions and ovaries were not seen separately [Fig.1].



**Fig 1:** Axial CECT image shows large bilateral adnexal masses (asterisks), having multilocular cystic appearance with septations and solid component (arrows).

The clinical and radiological findings were suggestive of neoplastic (likely malignant) bilateral ovarian masses. The patient was taken up for staging laparotomy. Peroperatively, uterus was found adhered to the right adnexa and both ovaries were found to be enlarged and unhealthy. Total abdominal hysterectomy with bilateral salpingo-oophorectomy, omentectomy and bilateral

pelvic lymphadenectomy was performed. On gross examination, right ovary was enlarged, multilobulated with intact capsule measuring 24x16x8cm. On cut it was solid cystic, multiloculated and was filled with thick yellow inspissated material along with areas of hemorrhage and necrosis [Fig.2a, 2b].

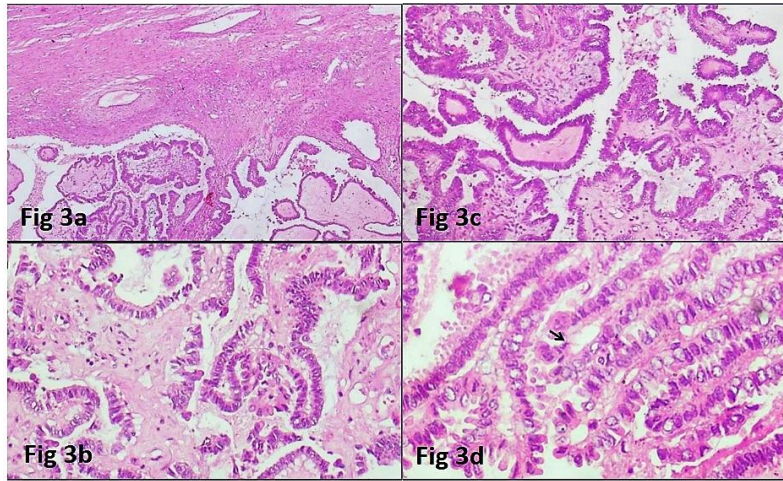


**Fig. 2(a):** Gross photograph shows a multilobulated right adnexal mass measuring 24x16x8cm with intact capsule. **(b):** Cut surface shows a multiloculated cystic mass filled with inspissated material along with few gray white solid areas and papillary excrescences.

Gray white, firm solid areas with friable papillary projections could also be seen. Left ovary was solid, gray white and measured 2.5x1.5x1cm.

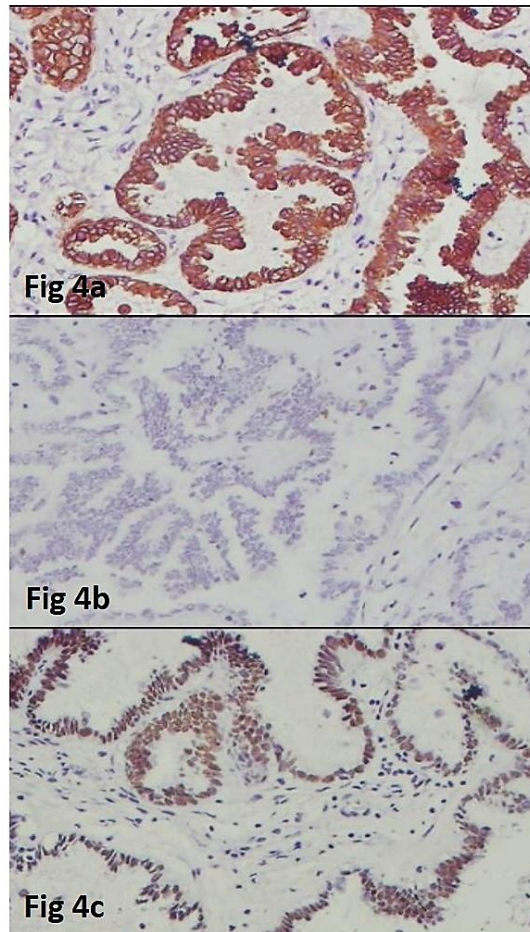
Hematoxylin & eosin (H&E) stained sections from the right ovarian mass showed a solid cystic tumour comprising of cells arranged in papillary pattern and nests. The papillae were broad based arranged in a hierarchical branching pattern, had a central fibrovascular core and were lined by atypical tumor cells. The tumor cells showed mild

pleomorphism and atypia. The individual cells were low cuboidal to columnar, with moderate amount of eosinophilic cytoplasm and round to oval hyperchromatic enlarged nuclei. Stromal invasion measuring more than 3 mm in depth in the form of tumor glands and nests could be seen with occasional atypical mitoses. Compressed ovarian stroma could be seen in the periphery and there was no capsular breach. [Fig 3a, 3b, 3c, 3d]



**Fig. 3(a):** H&E section shows a cystic ovarian tumor lined by cells arranged in broad based papillae with heirarchial branching pattern.(b): Tumor cells are invading into ovarian stroma in the form of papillae, glands and small nests.(c): Tumor cells show minimal stratification along with mild pleomorphism and nuclear atypia suggestive of low grade morphology.(d): Higher magnification shows cuboidal to columnar epithelial cells having moderate amount of eosinophilic cytoplasm, round hyperchromatic nucleus with single conspicuous nucleolus. A single atypical mitosis seen (arrow)

Tumour cells showed positivity for CK 7 and WT1, while negativity for CK 20. [Fig 4a, 4b, 4c]



**Figure 4:** Tumor cells are (a) CK 7 positive (b) CK 20 negative and (c) WT1 positive (x200).

Left ovary, uterus, bilateral fallopian tubes, omentum and the resected pelvic lymph nodes were free of tumour. Final diagnosis of grade 1 serous cyst adenocarcinoma right ovary, pT1aN0 (stage Ia) was offered. The patient was advised to be on close follow up with serum CA 125 levels. She is free of tumor 9 months after surgery.

## DISCUSSION

Serous ovarian tumours comprise 30% of all the ovarian tumours and nearly 50% of all the malignant ovarian tumours with 30-50% bilaterality. [1] Of all the serous tumours, 35% are invasive carcinomas. Patients usually present with abdominal pain and distension due to ascites or due to abdominal mass. It usually occurs in elderly females with a peak incidence in 60-70 years.

Incidence of malignant ovarian tumors in pediatric and adolescent age group varies from 10-35%, most of them being germ cell tumors. Though the reported incidence of surface epithelial ovarian carcinoma in adolescent age group varies from 0.5-1.5%, data on the clinical and prognostic features of these tumors in females younger than 30 years of age is not clear. [1-5] In a retrospective study on 71 Chinese patients of ovarian carcinoma under the age of 35 years, 40 cases (56.3%) were of serous cystadenocarcinoma and 22 cases (30.9%) were mucinous cystadenocarcinoma, and all of these carried a good prognosis. [3] In an Indian study, of the 151 girls under 20 years of age, thirty four (22.5%) turned out as malignant. Of these, nine (5.9%) were diagnosed with malignant surface epithelial tumor- mucinous cystadenocarcinoma (6 cases-3.9%) and serous cystadenocarcinoma (3 cases-1.9%). Malignant germ cell tumors were the most common (22 cases) accounting for 39% of malignant tumors.[4] The relatively higher incidence of malignant surface epithelial tumor (5.9%) in their study was attributed to the rural population and inclusion of cases up to 20 years of age. Surprisingly, the incidence of mucinous carcinoma was higher than serous in their study, which is contrary to the existing literature.

Recently two tier system of grading serous cystadenocarcinomas has been introduced dividing tumors into low grade and high grade. Low-grade tumors are considered slow growing and usually present at an early stage, as seen in our case. These patients are younger with a prolonged survival and

excellent prognosis as compared to those with high grade morphology. [3, 6, 7] The recommended treatment protocol for low grade stage I tumours is mainly surgical resection without the need for adjuvant therapy, which includes cystectomy with preservation of ovary or unilateral salpingoopherectomy. [3,7,8] In view of clinicoradiological suspicion of bilateral malignant ovarian tumor and adhered ovaries peroperatively, fertility preservation was not possible in our patient. Surgery and adjuvant platinum based chemotherapy are the mainstay of treatment for high grade ovarian carcinoma. Hyperthermic intraperitoneal chemotherapy (HIPEC), hormone therapy and targeted therapy like bevacizumab are other novel treatment options for higher grade and advanced stage ovarian surface epithelial tumors. [9]

CA 125 is a tumor marker which is a useful preoperative test to differentiate malignant surface epithelial ovarian neoplasms especially the serous carcinoma. Elevated levels (>35 U/ml) are found in more than 80% of epithelial ovarian carcinoma. Progressive fall or rise in the serum values can depict course of the disease during follow up. Hence it is an important aid to diagnosis and prognosis in equivocal cases. [10]

Germ cell tumors (GCT) are the most common ovarian tumors, accounting for 70% of all ovarian neoplasms in patients younger than 30 years of age. [4] These neoplasms present as solid cystic ovarian mass and hence remain an important preoperative differential diagnosis in this age group. Radiologically malignant GCTs appear predominantly as mixed solid cystic masses, thus can be difficult to differentiate from the more common malignant epithelial neoplasms. Role of tumor markers especially elevated alkaline phosphatase, lactate dehydrogenase, alpha fetoprotein and hCG levels in the diagnosis of GCT cannot be overemphasized. CT and MRI are more accurate, enable pre-operative extent delineation and may help arrive at a reasonable diagnosis together with the clinical details and tumor markers. [10]

To conclude, surface epithelial carcinoma is very rare in women younger than 30 years of age with a reported incidence varying from 0.5-5.9% in various studies, with the nadir in premenarchal age group. Serous cystadenocarcinoma as the most common surface epithelial carcinoma followed by

mucinous. [3] SC is usually unilateral, grade I and presents at an early stage with extremely good prognosis, risk of recurrence and metastasis being very low in this age group. Hence fertility preserving conservative management can be done in young and nulliparous females. Though unusual, gynaecologists and pathologists should be aware of the differential diagnoses, treatment options and

excellent prognosis of ovarian serous cystadenocarcinoma in patients younger than 25 years of age.

**Conflict of Interest:** Nil

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