

Case of Squamous Cell Carcinoma Transformation in Ovarian Mature Cystic Teratoma: A Case Report

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ABSTRACT

Mature cystic teratoma (MCT) is the commonest benign tumour arising in an ovary. Malignant transformation in a MCT of ovary is exceptionally rare, seen only in 1-2% cases. The most common malignant transformation in a benign teratoma of ovary is squamous cell carcinoma. The other malignant transformations in teratoma include adenocarcinoma, melanoma and basal cell carcinoma. The patients having this malignant transformation in teratoma have a very low 5-year survival rate of 20% approximately. Here, we describe a case of advanced-stage squamous cell carcinoma arising in a mature cystic teratoma in a 34-year-old woman. This solid high grade malignancy was missed in the pre-operative imaging which showed a right adnexal mass with echogenic foci within and septations, and was reported as a dermoid cyst. The intraoperative frozen section too taken from the thin portion of the cyst wall was reported as cystic teratoma. However in the staging laparotomy, right salpingo-oophorectomy and left oophorectomy, specimen the patient was reported to have moderately differentiated squamous cell carcinoma arising in a benign mature cystic teratoma with extensive tumour necrosis with omental tumour deposits. In view of initial incomplete surgery and advanced stage, decision of completion cytoreductive surgery was taken. The patients with mature cystic teratoma with high age (post-menopausal), a size of more than 10 cm and increased serum tumour markers should be suspected to have a malignant transformation and should be investigated thoroughly.

Keywords: Ovarian Teratoma, Squamous Cell Carcinoma

Introduction

Ovarian mature cystic teratoma (MCT), is a benign tumour composed of elements from all the three germ cell layers which include endoderm, mesoderm and ectoderm. This tumour accounts for 15% of all ovarian tumours in women between the ages 20-40 years. Only approximately 2% of these tumours undergo a malignant transformation. Out of these malignant transformed MCT approximately 70% are squamous cell carcinomas (SCC). The other rarer malignancies in MCT include adenocarcinoma, malignant melanoma and basal cell carcinoma.^[1] In this case report, we present a case of MCT showing a squamous cell carcinoma in an advanced stage which was missed during the clinical examination and the pre-operative imaging.

Case report

A 34-year-old woman presented with a four months history of progressing dull aching pain and gradual abdominal distension. Per abdomen examination revealed a 24 weeks equivalent sized well defined solid-cystic mobile mass, originating from pelvis. There were no signs of ascites. On per speculum and per vaginum examination, the cervix/vagina were healthy, uterus was pulled up and felt separate from the mass. On per rectum examination, the rectal mucosa and Pouch of Douglas (POD) was free. The serum

tumour markers were not raised: CA-125- 31.6 U/ml, CEA - 0.7 ng/ml, CA19.9 - 0.2 U/ml, β HCG - 0.1 mIU/ml and AFP - 0.1 ng/L. On ultrasonography, a complex adnexal mass measuring 10x12x11 cm with echogenic foci within and septations was noted which was reported as dermoid cyst. CT scan of abdomen and pelvis revealed mass with well-defined capsule in the left adnexa. The imaging showed the growth to have a mixed echogenic pattern consisting of cystic and solid components, which was finally reported as a mature teratoma of ovary (Fig 1a,b).

Intra-operatively the right ovarian solid cystic mass was removed. The frozen section from the cyst wall was reported as a mature cystic teratoma. On gross examination the mass measured 14 x 14 x 10 cm, had a smooth greyish external surface with few hemorrhagic areas (Fig 1 c). On cut surface the cyst was filled with yellowish white pultaceous material and tuft of hair. Few firm to hard areas were also noted (Fig 1d). The other specimens included biopsy from left ovary, mesentery, omentum, right paracolic peritoneum, left paracolic peritoneum, POD, bladder peritoneum and appendices epiploicae.

Histopathological examination showed the cyst lined by highly dysplastic keratinized stratified squamous epithelium, invading the underlying stroma in form of nests

and chords (Fig 2a). Tumour cells were large polygonal with high N:C ratio, pleomorphic hyperchromatic to vesicular nuclei. Individual cell keratinization and occasional keratin pearls were seen (Fig 2b). Tumour showed extensive necrosis along with brisk mitosis. Tumour also showed presence of hair follicles and adipocytes, however no immature or neuroepithelial component was noted (Fig 2c). Omental biopsy showed tumour deposits of squamous cell carcinoma (Fig 2d). All other specimens were free of tumour deposits. The patient was staged IIIA (TNM pT3aNxMx) as per International Federation of Gynaecology and Obstetrics (FIGO) classification. In view of initial incomplete surgery and advanced stage, a total abdominal hysterectomy along with left salpingo-oophorectomy, supracolic omentectomy and resection/anastomosis of ileal segment was done. The patient then received adjuvant chemotherapy and underwent six cycles of combination chemotherapy with paclitaxel+ carboplatin. Patient is disease free as yet. In view of rarity of this malignancy, no standard chemotherapy has been recommended by NCCN guidelines.

Discussion

SCC arising in ovary can arise either due to malignant transformation of a benign teratoma or due to a metaplastic

squamous change in endometriosis undergoing a malignant change. Pure de-novo SCC of ovary is rarely reported.^[2] SCC arising from a MCT in most instances is not diagnosed preoperatively akin to our case. No specific clinical signs and symptoms designate a MCT to have undergone a malignant transformation. In this index case the lady had a history of abdominal pain and distension for a period of four months. The literature reports these patients to have abdominal distension and bloating sensation along with the pelvic mass, similar to a benign MCT.^[3] The radiological diagnosis of a MCT is fairly simple as the components like bone, teeth and cartilage are easily identified. However, contrastingly the preoperative radiological detection of malignant transformation of MCT is very difficult.^[4] Hence there is a need to have objective ways to lay down the risk assessment of MCT having chances of a malignancy, as it is a very common tumour. These indirect evidences which signify a malignancy in MCT include: -high age (>50yrs), tumour size (>10cm), and elevated serum tumour markers.^[5]

Most of the times a SCC transformation in MCT is generally reported in relatively older post-menopausal ladies; however sometimes the condition is seen in younger

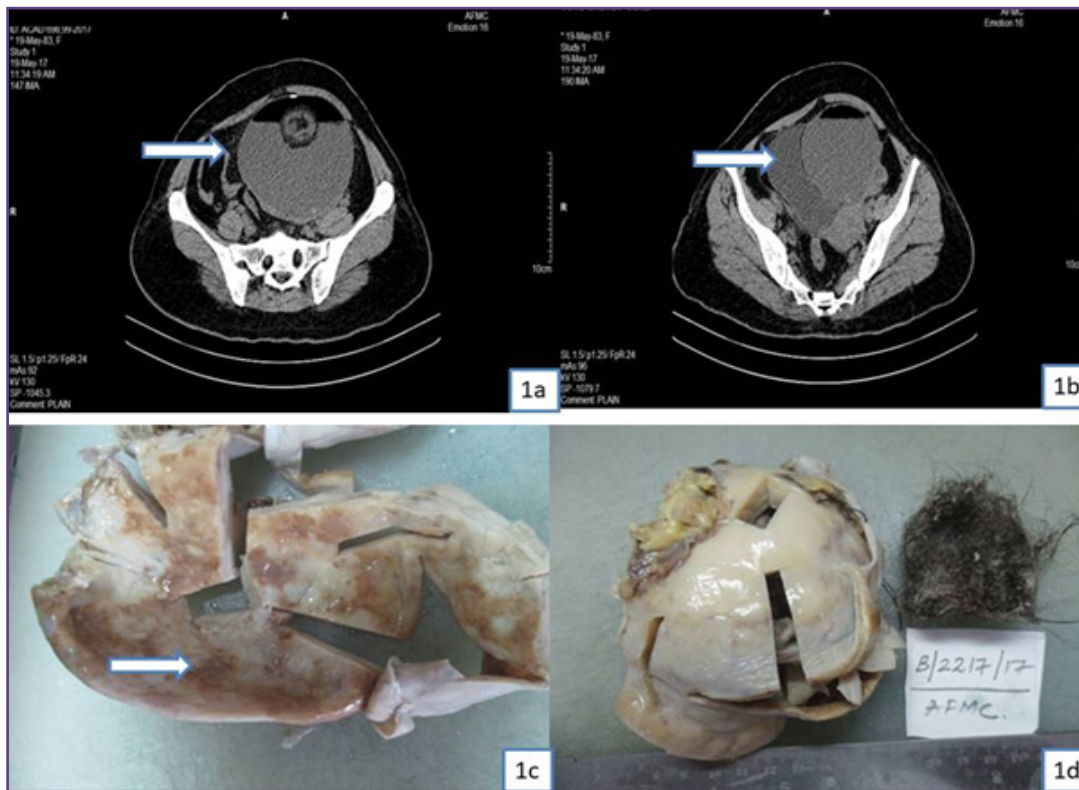


Fig. 1: (a&b) CT Scan abdomen and pelvis showing a well encapsulated mass in left adnexa; (c) cut surface gross showing a well encapsulated left adnexa mass greyish white in colour with few haemorrhagic areas; (d) external surface is smooth glistening- also seen is a tuft of hair taken out from the cavity.

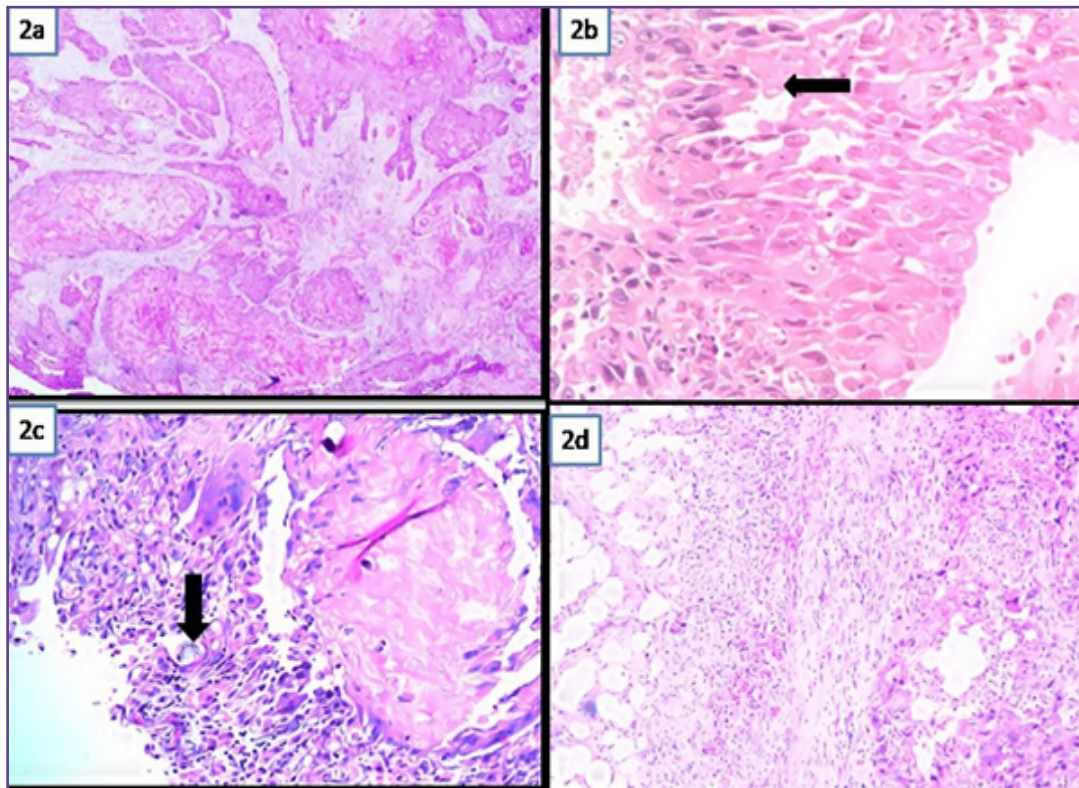


Fig. 2: (a) cyst lined by dysplastic keratinized stratified squamous epithelium,invading the underlying stroma in form of nests and chords (H&E x100); (b) cells have high N:C ratio and moderately pleomorphic hyperchromatic to vesicular nuclei. Individual cell keratinisation and occasional keratin pearls are seen(H&E x 400),(c) tumour showing malignant squamous cells and hair shaft(H&E x 200),(d) Omental biopsy showing tumour deposits in form of islands and clusters of atypical squamous cells(H&E x100).

group too.^[6] The patient's age in this reported case was 34 years, which is not in accordance to the reported literature. Study by Kikkawa et al reported that the chances of MCT are higher when patient's age is higher than 45yrs.^[7]

The research has shown that tumour size too has direct bearing on malignancy in MCT. In the case that has been discussed above, the maximum dimension of tumour was 12 cm, which is relatively larger than an average size benign ovarian teratoma cyst. A published case series by Kikkawa et al. concluded that tumours larger than 9.9 cm are more prone to develop malignancies with a sensitivity of 86%.^[7] Patients with malignancies in benign teratoma have a very grave prognosis with a 5 year survival of mere 20%.^[8] Other than the features discussed above the conventional prognostic markers of malignant MCT include: -FIGO stage,^[9]intra-peritoneal rupture of cyst, grade, lymphovascular invasion and the pattern of tumour infiltration (infiltrating/broad front).Patients of MCT with SCC as the malignant component fare better than the MCT which harbor adenocarcinoma, melanoma or basal

cell carcinoma¹.Peterson et al in a study of 190 malignant teratoma have reported a very high metastatic rate (64%).^[9]

Surgery forms the main stay of management of teratomas with malignancies.^[6,9] In nulliparous women especially in reproductive age group having early stage malignant teratoma, a simple conservative unilateral oophorectomy is offered. However, in the postmenopausal women, radical surgery like total abdominal hysterectomy with bilateral salpingo-oophorectomy is planned.^[6] Post-surgery combination chemotherapy, radiotherapy, or both are used. No large trials have been carried out in these sets of patients to have a consensus on chemotherapy/radiotherapy protocols.^[9,10] In conclusion, the gynecologist and pathologists should always entertain a differential of a malignancy in a teratoma when the patient is old, has an unusually large cyst and has raised serum tumour markers.

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