

DOI: <http://dx.doi.org/10.18203/2320-1770.ijrcog20174077>

Case Report

Ovarian tumor with mature teratoma with squamous differentiation in a postmenopausal female

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Received: 21 June 2017

Revised: 04 July 2017

Accepted: 19 July 2017

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ABSTRACT

Mature cystic ovarian teratomas are the commonest germ cell tumour accounting for about 30-45% of all ovarian neoplasms and around 60% of all benign tumors arising in the ovary. The reported incidence of malignant transformation in MCT is approximately 2%. A rare case of squamous cell carcinoma arising in a dermoid cyst of the ovary presenting in a postmenopausal female is presented here. A 60-year-old postmenopausal lady, presented with the complaint of pain in lower abdomen for 2 years and a large complex abdomino-pelvic mass on examination and investigations. Final histopathology was reported as squamous cell carcinoma arising from mature cystic teratoma of right ovary. Primary cytoreductive surgery was done and a surgical stage IIB was assigned. In view of the poor prognosis, adjuvant chemotherapy was started.

Keywords: Dermoid cyst, Mature cystic teratoma, Squamous cell carcinoma of ovary

INTRODUCTION

Mature cystic ovarian teratomas (a more appropriate term than the commonly used “dermoid cysts”) are the commonest germ cell tumour found predominantly in young women with a median age of 30 years.¹ It accounts for 20% of all ovarian neoplasm.² These tumors are composed of well-differentiated derivations from at least two of the three germ cell layers (ectoderm, mesoderm, and endoderm). Clinically, it is usually asymptomatic but may be discovered accidentally during gynaecologic investigations for other conditions or due to mass effect.² When it is discovered during childhood, ovarian cystectomy is curative.

However, if discovered in women above 50 years of age, the likelihood of malignant transformation of any of the components is very strong.³ Mature cystic teratomas are

the most common ovarian neoplasm removed at surgery.^{4,5} The tumors are bilateral in about 10% of cases.⁶ The gross pathologic appearance of mature cystic teratomas is characteristic. The tumors are unilocular in 88% of cases and are filled with sebaceous material, which is liquid at body temperature and semisolid at room temperature.⁶ Squamous epithelium lines the wall of the cyst, and compressed, often hyalinized ovarian stroma covers the external surface.^{7,8}

Hair follicles, skin glands, muscle, and other tissues lie within the wall. There is usually a raised protuberance projecting into the cyst cavity known as the Rokitansky nodule. Most of the hair typically arises from this protuberance. When bone or teeth are present, they tend to be located within this nodule.⁹ Ectodermal tissue (skin derivatives and neural tissue) is invariably present.^{6,7,9} Mesodermal tissue (fat, bone, cartilage, muscle) is present in over 90% of cases, and endodermal tissue (eg,

gastrointestinal and bronchial epithelium, thyroid tissue) is seen in the majority of cases.⁶ Adipose tissue is present in 67%-75% of cases, and teeth are seen in 31%.^{6,7,9} Mature cystic teratomas (MCT; also called dermoid cysts) account for about 30-45% of all ovarian neoplasms and around 60% of all benign tumors arising in the ovary with a 0.17-1.4% reported incidence of malignant transformation.¹⁰⁻¹² Squamous cell carcinoma in MCTs is most commonly seen in postmenopausal women.³ There are no definitive clinical features, tumor markers are not often raised and imaging methods are many times not helpful. Hence, most cases are diagnosed postoperatively.¹³ Tumors confined to the ovary usually have a better prognosis and patients with stage III or IV disease rarely survive five years.¹⁴

Here, we are presenting a case of squamous cell carcinoma of the ovary which was diagnosed at an early stage and was managed appropriately keeping in mind the poor prognosis of the condition.

CASE REPORT

A 60-year-old postmenopausal lady, para 4 live 4, postmenopausal for 15 years, presented in the department of Obstetrics and Gynaecology, Safdarjung Hospital, New Delhi with the complaint of pain in lower abdomen for 2 years. Her general condition was good. There was no visible or palpable evidence of any neck swelling, breast abnormality or lymphadenopathy. Her per abdominal examination showed a midline, large 14-16 weeks size, firm to hard (variegated), non-tender, well-defined, freely mobile mass arising out of pelvis. Her pelvic exam revealed a mass on right side of a normal sized uterus, stuck to uterus, with loss of intervening space and moving with cervical movement. Her rectal examination ruled out any POD nodularity. Routine and specific investigations were carried out. USG whole abdomen demonstrated a large cystic (107*102*80 mm) mass arising from pelvis and occupying lower abdomen. CECT of the abdomen reported a large thin walled cyst showing fat fluid level with heterogenous enhancing solid component at the base of lesion-likely ovarian dermoid with? Neoplastic transformation. Tumor markers were carried out and values were reported as follows:

- CA 125: 51.3 U/ml
- CEA: 24.7ng/ml
- AFP: 3.0 U/ml.

In view of clinical, radiological and biochemical findings, patient was prepared for staging laparotomy with facility of frozen section per-op.

Her intraoperative finding revealed a large 10*10cm right sided ovarian mass. The mass was densely adherent to omentum, intestine, pelvic peritoneum and sigmoid colon. Adhesiolysis was done and the mass after separation was sent for frozen section reporting. Meanwhile total abdominal hysterectomy with

contralateral salpingo-oophorectomy was completed. Frozen section report suggested possibility of immature teratoma (malignant). Complete surgical staging and primary cytoreduction was done. Deposit over sigmoid colon was removed.

Final histopathology was reported as squamous cell carcinoma of right ovary arising from dermoid cyst. Both fallopian tubes, parametrial tissues, peritoneal biopsies, uterus, cervix, omentum and pelvic lymph nodes were unremarkable. Deposit from sigmoid colon showed deposit from the tumor. Peritoneal fluid was positive for malignant cells.

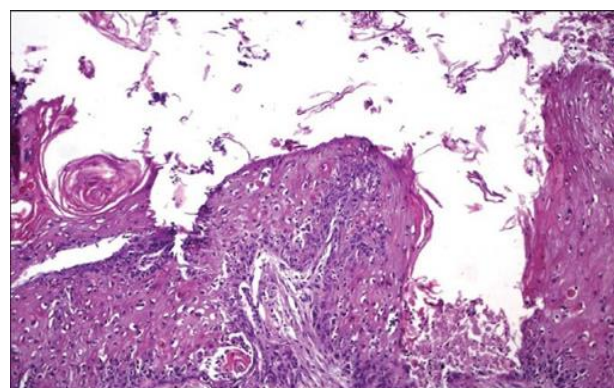


Figure 1: Histopathological picture of MCT.

For management, the patient was assigned to squamous cell carcinoma of the ovary arising in a MCT, surgical stage IIB. Her postoperative period was uneventful. In view of poor prognosis, adjuvant chemotherapy (six cycles of BEP regimen containing Paclitaxel and Carboplatin) was started.

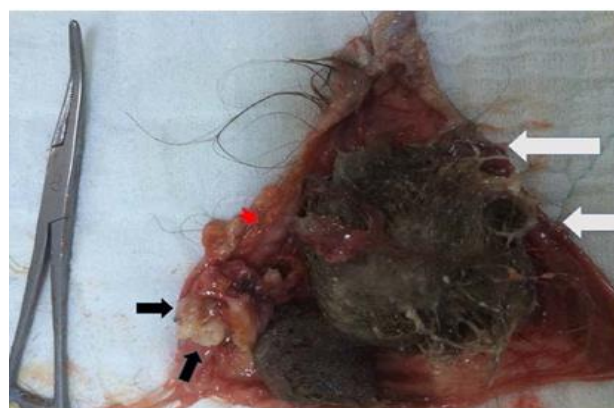


Figure 2: Gross view (cut section) of MCT.

DISCUSSION

Mature cystic teratoma (MCT) is a common ovarian neoplasm.^{15,16} Mature cystic teratoma is the most prevalent germ cell tumor in women of reproductive age. Malignant transformation of an MCT is an uncommon event occurring in less than 2% of all mature dermoid

cysts.¹⁷ Malignant transformation starts at the postmenopausal period with an average age of 55 years; the most common malignant transformation occurs to squamous cell carcinoma.¹⁸ Other malignancies include adenocarcinoma, sarcoma, carcinoid, thyroid carcinoma and melanoma.¹⁹ Non-teratomatous squamous cell carcinoma of the ovary has been found to originate from endometriosis, epidermoid cysts or from the surface epithelium.¹⁸ Age and size are predictors of malignant transformation in benign teratoma. Malignant transformation tends to occur more commonly in older women with an average mean age at diagnosis of 55 years.¹⁷ Malignant transformed mature cystic teratomas have a larger size (mean size of 15 cm) compared with MCTs (mean size of 6-9 cm).¹⁷

The malignant transformation usually begins from the "dermoid plug".²⁰ Since the malignant transformation is rare and the malignant form resembles the appearance of the benign form, it is challenging to make a preoperative diagnosis. Malignant transformation usually occurs after menopause and when it occurs, the prognosis is grave.⁴ Rim et al reported that the malignant transformation occurred at an average age of 56.8 years, and 63% of the cases were older than 40 years old at the time of diagnosis.⁵ Malkasian et al reported that in a 612-case series the malignancy risk was doubled in patients over 50 years of age.²¹

Symptoms at presentation are variable in both diagnoses with some patients presenting with acute abdominal pain and others with constitutional symptoms such as fatigue, urinary symptoms and anorexia/weight loss.²⁰ Although tumor markers may be raised in patients with squamous cell carcinoma arising from MCT, it is difficult to use tumor markers to distinguish between MCT and squamous cell carcinoma arising from an MCT since tumor markers can also be elevated with MCT.⁴ For example, squamous cell antigen (SCC) levels were

found to be significantly higher in patients with squamous cell carcinoma arising from MCT than with MCT alone.⁵ However, mean levels in squamous cell carcinomas are lower than in patients with adenocarcinomas, and cannot predict the diagnosis preoperatively.⁵ CA19-9 is another tumor marker found to be significantly higher in patients with squamous cell carcinoma arising from MCT than with MCT alone, however it is also a difficult marker to use in preoperative screening since the mean levels of CA 19-9 are found to be elevated in patients with MCT alone.⁵

Yeol Park et al. reported that age at the time of diagnosis and tumor size performed well than levels of CA125 or CA19-9 in detection of malignant transformation in mature cystic teratomas. They indicated that 45 years of age as the patient age and 99 mm as the tumor size were the optimal cut offs. Malignant ovarian tumors may sometimes show an elevated level of SCC, the marker for squamous cell carcinoma.²²

Radiologically, mature teratomas may demonstrate a broad spectrum of findings ranging from a purely cystic mass, a fat-containing mass or a heterogeneous soft tissue mass. Classic diagnostic findings for MCT include fat attenuation within a cyst that possibly also contains calcification.²¹ Imaging features concerning for malignant transformation include thick walls, enhancing solid components or papillary projections within the cyst, peritoneal deposits or lymphadenopathy. In our case the availability of prior CT imaging demonstrating a fat-containing adnexal mass with areas of calcification and a subsequent CT that demonstrated interval enlargement with a new, enhancing soft tissue component was concerning for malignant transformation of the MCT.

MRI may be helpful. CEA may be elevated. Other rare pathologies in MCT are adenocarcinoma and melanoma. Takagi et al have found CEA to be more useful than CA 125 and CA 19-9 in malignant transformation of MCT.¹⁶ This entity carries very poor prognosis especially when diagnosed in advanced stage. Optimal cytoreduction is difficult to achieve in advanced disease.

Suspicious features (clinical and on MRI) include

- Postmenopausal women (mean age-55years vs. 37.5 years for benign dermoid cysts).
- Larger size of tumor (mean- 152mm vs. 88 mm for benign).
- Solid areas within the cyst.
- Invasion of adjacent organs/capsule.^{14,23}

Given the rarity of squamous cell carcinoma of the ovary, there is no clear standard of care and often patients are treated similar to patients with epithelial ovarian cancer.¹⁹ MCT can be managed expectantly in premenopausal women with ovarian dermoid cysts less than 6 cm in diameter, especially if pregnancy is desired. The mean growth rate of dermoid cysts in premenopausal women is 1.8 mm/year.²² However, if tumor markers are abnormal and worrisome imaging findings are noted, a surgical approach is advised.⁴ Ovarian torsion and rupture are other complications of MCT to consider in addition to malignant degeneration.²¹ Masses such as MCT that can predispose the ovary to torsion can also mimic torsion.²⁴ The most common imaging finding of ovarian torsion is ovarian enlargement and cross-sectional imaging may show lack of enhancement.²⁴ Enhancing solid components and/or papillary projections would be less characteristic of ovarian torsion and rather of malignant degeneration.

Review of multiple case series recommends surgery including total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy and lymph node dissection.^{18,19} Standard treatment includes chemotherapy with a platinum-based agent. The role of radiotherapy remains undefined.^{18,19} The stage of disease is the best predictor of survival with overall better outcome for

patients with limited disease with disease-free survival for up to 5 years after diagnosis.

A systematic review and analysis of published data was done by Hackethal et al.³ Sixty-four suitable studies provided information on 277 patients. It was observed that squamous cell carcinoma in MCT was mainly found in women above 50 years of age, having a high concentration of CA 125 and ovarian tumors more than 10 cm in size. They also found that FIGO stage Ia had better survival than those with advanced disease. Complete resection with advanced disease followed by adjuvant chemotherapy was seen to be associated with higher survival. According to them adjuvant radiotherapy did not improve survival.

A case series and review of literature was published in European Oncology and Hematology, 2013.²⁵ In this retrospective review conducted over 24 years between 1986 and 2010, they found six women treated for squamous cell carcinoma in MCT, all stage III/ IV. They found that durable responses were difficult to achieve but best treatment response was seen in a woman who had partial response to chemo-radiotherapy (survival 19 months). According to them, concurrent chemo-radiation could be considered for disease confined to pelvis. However, median survival in their series was only 12.5 months.

Sporadic case reports have been published by Indian authors on this subject.^{26,27} These reports are similar to our case. However, Khajuria et al and Santwani et al have reported squamous cell carcinoma in dermoid cyst at younger ages i.e., 37 years and 40 years, respectively.^{28,29}

CONCLUSION

Keeping in view the rarity and poor prognosis of squamous cell carcinoma of the ovary arising in a mature cystic teratoma, it is very essential for a gynecologic oncologist to be aware of this condition and be equipped to deal with it.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Rai A, Zutshi V. Ovarian tumor with mature teratoma with squamous differentiation in a postmenopausal female. *Int J Reprod Contracept Obstet Gynecol* 2017;6:4135-9.