**Squamous Cell Carcinoma Arising in a Mature Cystic Teratoma of the Ovary: A Case Report**

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**ABSTRACT**

Mature cystic teratomas account for about 30-45% of all ovarian tumors. Malignant transformation in these tumors is a rare event, occurring in only 0.17-2% of cases, with squamous cell carcinoma being the most common type. As there are no specific signs and symptoms to suggest malignancy in dermoid cyst, it is difficult to predict, and most cases are diagnosed postoperatively. We report a 55-year-old postmenopausal lady who presented with the complaint of pain in abdomen and abdominal distension for four months and a large complex abdomino-pelvic mass evident clinically and radiologically. Final histopathology was reported as malignant transformation in mature cystic teratoma. The patient was assigned to squamous cell carcinoma of the ovary arising in a mature cystic teratoma, surgical stage IIC. In view of the poor prognosis, adjuvant chemotherapy was given.

Keywords: dermoid, malignant transformation, mature cystic teratoma, squamous cell carcinoma.

**Introduction**

Mature cystic teratomas (ovarian dermoid) account for about 30-45% of all ovarian neoplasms and around 60% of all benign tumors arising in the ovary.¹ Malignant transformation of the various mature tissue components of a dermoid cyst is rare and the reported incidence is 0.17-1.4%.²⁻³ Squamous cell carcinoma (SCC) is the commonest form of malignant transformation accounting for >80% of cases, followed by adenocarcinomas and carcinoid tumors.⁴ SCC in mature cystic teratomas is most commonly seen in postmenopausal women.⁴ There are no distinctive clinical features, tumor markers are often normal and pre-operative radiological diagnosis is difficult. Hence, most cases are diagnosed post-operatively.⁵ Tumors confined to the ovary have a better prognosis and patients with stage III and IV disease rarely survive for five years.⁶ At the time of presentation, the most frequent symptom associated with malignant transformation is lower abdominal or pelvic pain and increasing abdominal girth.⁷ Old age, large tumor size, and solid portion in mature cystic teratoma seem to predict the malignant transformation.⁵,⁶ We describe such a rare case of squamous cell carcinoma arising in a mature cystic teratoma with review of related literature.

**Case Report**

A 55-year-old postmenopausal lady, grand multipara, presented in gynecologic oncology unit of Civil Service Hospital, Kathmandu, Nepal with the complaint of pain in abdomen and abdominal distension for four months and unable to pass flatus and urine for one day. She reported one episode of per vaginal bleeding for two days. She had no known medical history or family history of cancer. There was no visible or palpable evidence of any neck swelling, breast abnormality or lymphadenopathy. On physical examination, her abdomen was grossly distended and tensed making it difficult for assessment of mass.

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On ultrasonography a cystic lesion of around 10.4 × 9.5 × 8.8 cm was noted to the right side of the midline with solid and hyperechoic contents within the cyst with increased vascularity suggestive of dermoid cyst or malignant tumor of ovary. CT scan with contrast showed large cystic pelvic mass measuring 12 × 11 × 8.5 cm with fat along with calcifications and heterogeneously enhancing solid components with possible origin from left ovary, multiple heterogenous enhancing soft tissue density lesion in periphery largest measuring 6.3 × 6.2 × 4.5 cm and intraperitoneal fluid collections with multiple fat fluid levels (Figure 1). Additional findings were multiple enhancing bilateral para-aortic lymph nodes and mild left hydroureteronephrosis suggestive of immature teratoma. Serum tumor markers were as follows: CA-125: 71.7 U/ml and CEA: 5.53 ng/ml.

Figure 1: CT scan of the abdomen and pelvis in coronal and axial view showing large cystic lesion with solid component and fat filled fluid level in the center of abdomen

Figure 2: Intraoperative picture showing ascites, tumour arising from left ovary, occupying Pouch of Douglous

Figure 3: Gross specimen showing tumour arising from left ovary, with cheesy material, tuft of hair and fleshy tissue.

Figure 4: A dermoid cyst showing area of squamous cell carcinoma
The patient underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy, omentectomy and para-aortic lymph node sampling. Gross examination showed ascites with dirty colored fluid about three liters with fat globules. Cystic mass measuring 12 × 10 cm on right and 6 × 6 cm on left with irregular firm growth over surface was noted which was densely adhered to omentum, sigmoid colon and rectum (Figure 2). Tumor deposits measuring 2 × 2 cm and 1 × 1 cm noted in omentum and small bowel respectively (Figure 2). On cut section revealed multiloculated cyst, filled with grumous material, hair tooth and fleshy mass. Intraoperative finding suggested stage IIIC ovarian cancer. Final pathological evaluation revealed malignant transformation in mature cystic teratoma in bilateral ovaries, omentum and para-aortic lymph nodes were free. Consequently, tumor stage was determined as stage IIC disease. Post-operative period was uneventful, and patient was planned for adjuvant chemotherapy.

Discussion

Mature cystic teratoma of the ovary, often referred to as dermoid cysts are benign and are one of the commonest ovarian tumors. They occur principally in premenopausal women with mean age at diagnosis of 35 years. Malignant transformation of these tumors is rare, occurs in 0.17-1.4% and are most commonly seen in postmenopausal women. Abdominal pain followed by abdominal or pelvic mass is the most common symptom which may be present in benign cystic lesions. As the tumor increases in size patient can present with compressing symptoms like gastrointestinal symptoms diarrhea/constipation, rectal bleeding or urinary frequency. Our patient had complained of abdominal pain, distension associated with pressure symptoms and one episode of per vaginal bleeding. Preoperative risk assessment for malignancy is extremely difficult but very important for treatment planning.

The risk factors for malignant transformation include age more than 45 years, tumor size >10 cm, rapid growth, imaging characters and increased tumor markers. James R Powell et al., in their study observed that mean age of malignant transformation in dermoid cyst was 55 years. Kikkawa et al., in their case series found that tumor diameter larger than 9.9 cm was 86% sensitive for malignant change. Importance of tumor markers is studied in many studies. According to Kikkawa et al., it was found that CEA was the best screening marker followed by SCC Antigen, both being superior to CA-125 and CA-19-9. Thus, measurement of CEA and SCC Ag levels in patients aged 45 years or older, who have dermoid cyst like ovarian tumor >9.9 cm was finally recommended that would provide a good clinical strategy for preoperative risk assessment in diagnosis of malignant change. In yet, another study Mori et al. reported that age >40 years and serum SCC Ag >2.5 ng/ml were 77% sensitive and 96% specific for malignant transformation which has also been useful in monitoring for recurrent disease. In our case, the age of the patient was 55 years, tumor diameter was 12 cm and tumor marker CEA (5.53 ng/ml) and CA-125 (71.7 U/ml) were elevated. Fewer reports exist regarding the radiological diagnosis of malignant transformation developing from a mature cystic teratoma. On CT, it appears as a fat containing tumor with an enhancing, irregularly marginalized solid component with areas of invasion through teratoma. The contrast enhancement of the Rokitansky protuberance should raise the suspicion of malignant transformation. MRI features include solid component with contrast enhancement, transmural or trans-septal extension, evidence of adhesion to surrounding structures, necrosis and hemorrhage. A recent case report has also demonstrated the value of MRI with fat suppression techniques to detect malignant transformation developing within a mature cystic teratoma.

It has been thought that squamous cell carcinoma in mature cystic teratoma arises from metaplastic squamous epithelium. Furthermore, high-risk HPV infection has also been thought to be associated with ovarian squamous cell carcinoma. Mostly mature cystic teratomas are detected 15-20 years before they undergo secondary malignant transformation. Cytogenetic abnormalities might precede histological changes, and prolonged exposure to various carcinogens in the pelvic cavity might cause the malignant changes in mature tissue. Thus, it is more common in postmenopausal patients. Squamous cell carcinoma arises from ectodermal component of dermoid cyst. There are two theories for origin of SCC i.e. from epidermal (80%) and respiratory tract. Montgomery protuberance at the junction between teratoma and normal ovarian tissue is the area of maximum cellular activity from where malignancies usually arise which eventually grow and extend to neighboring organs. During histopathological examination, this area has to be clearly examined to rule out malignancy. The definitive therapy for this condition has not yet been established because of rarity of the condition and incidental nature of diagnosis. In stage I disease with postmenopausal women, total abdominal hysterectomy with bilateral salpingo-oophorectomy is procedure of choice. Unilateral oophorectomy may be considered in women wishing...
to preserve fertility. Stage II or III disease require optimal debulking followed by combination of chemotherapy and radiotherapy.\textsuperscript{5, 10, 11, 18} Meta-analysis of published data suggests that chemotherapy regimen containing alkylating agents might improve overall survival.\textsuperscript{19} Some studies have shown that remarkable responses have achieved with paclitaxel based regimen combined with pelvic radiation.\textsuperscript{11} These tumors carry poor prognosis with a five years survival rate of only 15-30%. Prognostic indicators include FIGO staging, tumor rupture, vascular invasion and infiltration of surrounding structures.\textsuperscript{6}

**Conclusion**

Squamous cell carcinoma arising in dermoid cyst is a rare pathologic event with poor prognosis. Risk factors are over 45 years, tumor size more than 10 cm and increase serum tumor markers CEA and SCC Antigen. Tumors confined to the ovary usually have a better prognosis. Since, preoperative diagnosis of malignancy is very difficult, dermoid cysts in postmenopausal women should be adequately sampled to avoid an erroneous diagnosis. Awareness to this rare entity would help in early diagnosis and better management of older patients with dermoid cysts.

**References**