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Pathology Section

Squamous Cell Carcinoma Arising in a Dermoid Cyst Ovary: A Rare Case Report

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ABSTRACT

Dermoid cysts account for approximately 25% of all ovarian tumors. Presence of pure squamous cell carcinoma is very rare findings (1-2%), which is attributable to malignant transformations into a pre-existing dermoid cyst. As there are no specific signs and symptoms to suggest malignancy in dermoid cyst, therefore it is difficult to predict, and most cases are diagnosed post-operatively. We hereby report a case of dermoid cyst ovary with malignant

transformation into squamous cell carcinoma in 50 years post-menopausal women. Who presented with complaint of abdominal pain and increased frequency of micturation. Hysterectomy with Bilateral-salpingo-oophorectomy (BSO) with omentectomy was done. Right ovary showed huge cyst with hair shaft, sebaceous materials and a small solid area. Histopathological examination revealed squamous cell carcinoma was arising within the dermoid cyst.

Keywords: Malignant transformation, Ovarian tumor, Teratoma

CASE REPORT

A 50-year-old post menopausal woman presented with pain in abdomen with increased frequency of micturation since 1.5 years. She also developed off and on fever and vomiting for last 15 days. Finding of CT-scan was well defined peripherally enhancing cystic lesion in right ovary measuring 16.5x13.5x12.6 cm with heterogeneously enhancing soft tissue component with necrotic changes and fat fluid level extending from pelvis to umbilical region. Serum tumor markers: CA-125- 47.2 IU/ml, CEA- 14.3 ng/ml and beta-HCG was 7.85 mlU/ml. The clinical differential diagnosis was malignant ovarian tumor or malignant teratoma. Peroperatively tumor was fixed to bowel wall. Total abdominal hysterectomy with bilateral salpingo-oophorectomy with omentectomy (TAH BSO) was done. Gross examination showed a cyst measuring 17.0x14.0x12.0 cm. On cutting pultaceous material came out. Cut surface showed a bunch of hair and a gray white solid area measuring 6.0x5.5 cm with foci of necrosis [Table/Fig-1]. Microscopic examination showed cyst lined by stratified squamous epithelium with adenexal structures, keratin flakes and hair follicles [Table/ Fig-2 A and B]. Sections from solid area revealed malignant tumor (squamous cell carcinoma) arising from lining of cyst wall and proliferating in form of solid sheets and nests with comedonecrosis pattern. Tumor was moderately differentiated with occasional keratin pearls [Table/Fig-3 A and B]. P63 immunostaining showed diffuse nuclear

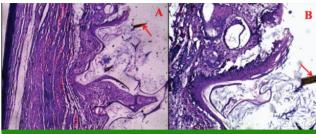


[Table/Fig-1]: Gross picture showing cut surface of dermoid cyst with bunch of hair and solid area (arrow) with foci of necrosis.

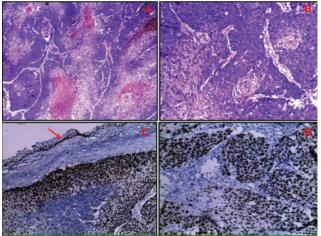
positivity in tumor cells [Table/Fig-3C and D]. Omentum showed positive peritoneal deposits. Post operative period was uneventful and patient was referred to medical oncology unit for further management, where adjuvant chemotherapy was started. Prognosis was very poor. After two weeks she developed ascitis and breathlessness and did not respond to medical treatment. Unfortunately patient lost the follow up.

DISCUSSION

Mature cystic teratoma also known as the dermoid cyst, which accounts for 25% of all ovarian tumors. It is usually found in young women during reproductive years [1]. These



[Table/Fig-2]: Cyst wall showing lining of squamous epithelium and acellular keratin and hair shaft (arrow) H&E \times 40X [A] and H&E \times 100X [B].



[Table/Fig-3]: Squamous cell carcinoma disposed in sheets and nests with comado-necrosis pattern H&E x 100X [A]. High power view (200X) [B]. Immunostaining p63 outlines the cyst wall (arrow) and below it tumor proliferation x100X [C]. Immunostaining p63 showing diffuse nuclear positivity in tumor cells x 200X [D].

tumors often grow slowly and go unnoticed till they become very large. About 1-2% of dermoid cyst undergo malignant transformation, are most commonly seen in postmenopausal women and associated with poor prognosis [1,2]. Mature cystic teratoma are unilocular/multilocular cysts contain hairs, sebaceous material and other tissues derived from embryonic germ layers which have potential to develop malignancy of divergent histomorphological types. The majority of such malignancies arising within ovarian dermoid are: squamous cell carcinoma (80%) followed by adenocarcinoma, carcinoid tumors and melanoma [3,4]. The denovo development of SCC in otherwise healthy ovary is extremely rare [5]. Preoperative diagnosis is difficult [6]. These tumors commonly spread through direct and local invasion rather than lymphatic or hematogenous route [7]. Clinical behavior of these tumors is unpredictable and the role of chemotherapy and radiotherapy remains unclear.

Most patients are asymptomatic or have symptoms of abdominal distension. As the tumor increases in size patient can present with compressing symptoms like gastrointestinal symptoms diarrhoea/constipation, rectal bleeding or urinary frequency [8]. Our patient had complained of abdominal pain and urinary frequency for 1.5 years.

Grossly in dermoid cyst there may be solid area having different types of tissue like cartilage, bone, soft tissue and hairs. If there is solid friable area with any sign of necrosis hemorrhage or irregular plague and thickened area in the cyst wall, then it should be sampled carefully to rule out immature component or malignant transformation in the cyst [1,4]. Histopathological examination plays an important role in confirmation of diagnosis and treatment planning. In our case there was typical histology of dermoid cyst but the solid area showed various stages of development from dysplasia to frank invasion (malignancy). Tumor cells were proliferated in the form of solid sheets and nests in desmoplastic stroma with occasional keratin peals and large area of necrosis. P63 was diffusely positive in tumor cells [Table/Fig-3C and D]. In our case differential diagnosis was malignant teratoma but we did not find other tumor component like sarcoma, immature neural component, melanoma or poorly differentiated tumor cells. Hence, the case was considered as pure squamous cell carcinoma arising in a dermoid cyst ovary.

The risk factors for malignant change in dermoid cyst are advance age more than 40 years, tumor size >10 cm, rapid growth and increased tumor markers [9,10]. James R Powell et al., in their study observed that mean age of malignant transformation in dermoid cyst was 55 years [11]. Kikkawa et al., in their case series found that tumor diameter larger than 9.9 cm was 86% sensitive for malignant change [10]. In our case the age of patient was 50 years, tumor diameter was 17 cm and tumor marker CEA (14.3 ng/ml) and CA-25(47.2 IU/ ml) were elevated. Mori et al., reported that age >40 years, serum SCC antigen >2.5ng/ml were 71% sensitive and 96% specific for malignant transformation which has also been useful in monitoring for recurrent disease [12]. Due to rarity of lesion as well as incidental nature of diagnosis, the definitive therapy for squamous cell carcinoma arising in a ovarian dermoid has not yet been established [13]. In stage-I disease with postmenopausal women total abdominal hysterectomy with bilateral salpingo-oophorectomy and omentectomy (TAH-BSO) is procedure of choice. While in younger patients who wish to preserve fertility, unilateral oophorctomy may be done. Patient with stage II or III disease required optimal debulking followed by combination of chemotherapy and radiotherapy [4,8,13,14]. JL Hurwitz et al., suggested that repeated surgical resection of disease at the time of relapse could give a very durable response in selected women. In their case series they observed no definite benefit from chemotherapy to any of their patients [14]. James R Powell in their case series used platinum based chemotherapy and out of six only two patient have partial response [11]. The prognosis for these tumors has often been reported to be poor with a five year survival rate of only 15-30% [2,9]. The potential predictors reported include FIGO stage, residual tumor, rupture or spillage, tumor grade, vascular involvement and the mode of tumor infiltration [10]. Meta-analysis of published data suggests that chemotherapy regimen containing alkylating agents might improve overall survival [4]. Some studies have shown that remarkable responses have achieved with paclitaxel based regimen combined with pelvic radiation [13].

CONCLUSION

Squamous cell carcinoma arising in dermoid cyst ovary is rare occurrence with poor outcome. Risk factors are age over 45 years, tumor diameter greater than 10 cm and increase serum tumor markers CEA and SCC antigen. Tumors confined to the ovary, usually have a better prognosis. Awareness to this rare entity would help in early diagnosis and better management of older patients with dermoid cysts.

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