VOLUME-7, ISSUE-5, MAY-2018 • PRINT ISSN No 2277 - 8160

Original Research Paper

Pathology

MALIGNANT TRANSFORMATION OF MATURE CYSTIC TERATOMA OVARY- A RARE OCCURRENCE

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ABSTRACT Mature cystic teratoma (MCT) also called dermoid cyst, is the most common ovarian neoplasm. Malignant transformation is rare and observed in 1-3% of all MCTs. Squamous cell carcinoma is the most common malignancy followed by adenocarcinoma. We describe a case of squamous cell carcinoma arising from a MCT in a 58 year post menopausal female. She presented with pain and heaviness in lower abdomen since two months. A large abdominopelvic mass was noted on local examination. The patient underwent hysterectomy with bilateral salpingo-oophorectomy. Final histopathology was reported as squamous cell carcinoma of left ovary arising from dermoid cyst and invading the left fallopian tube.

KEYWORDS : Dermoid; Squamous cell carcinoma; Mature cystic teratoma

In summary, preoperative diagnosis of ovarian SCC-MCT is challenging. Clinical, radiologic and intraoperative macroscopic findings are non-specific compared to those of dermoid cyst. A high level of suspicion is required in patients of advanced age with relatively large masses who are thought to have ovarian teratomas. Serum tumor markers might be a useful diagnostic tool to rule out SCC-MCTs. Awareness of this rare entity is required for early diagnosis and better management when dealing with older patients of mature teratoma ovary.

Introduction

Mature cystic teratoma (MCT) is the most common ovarian neoplasm and is composed of all three germ layer derivatives[1]. Majority of these occur in premenopausal women and carries a good prognosis. Malignant transformation is an uncommon complication and is observed in 1-2% of cases[1]. The most common malignancy is squamous cell carcinoma (SCC) (90%) followed by adenocarcinoma[2]. Majority of such patients are postmenopausal and the clinical outcome is poor[3]. Clinical features, imaging studies and serum markers have a limited role in its preoperative diagnosis. This case report aims to highlight such rare occurrence and this entity must be kept in mind when dealing with MCTs especially in older patients.

Case report

A 58 year old postmenopausal female presented to the outpatient Department of Dr BSA Medical College and Hospital with complaints of pain and heaviness in the left lower abdomen since two months. Local examination revealed a large firm to hard non tender abdominopelvic mass. In view of clinical suspicion of malignancy, Serum CA125 levels were done and reported as 37mIU. The patient underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy. Preoperatively, left ovary was enlarged which on cutting open was solid cystic with cheesy material and friable necrotic material. A presumptive differential diagnosis of degenerated dermoid ovary and carcinoma ovary was kept. Gross examination showed solid cystic left ovary measuring 9x8x6 cm with solid area measuring 6x5x3 cm (figure 1). Multiple sections examined showed nests of atypical squamous cells with large areas of necrosis (figure 2). A preliminary diagnosis of squamous cell carcinoma was made. However in view of rarity of primary SCC of ovary, extensive sampling was carried out which revealed a cyst lined by foamy macrophages, foreign body giant cells and occasional hair shaft (figure 3 & 4).

Final histopathology was reported as high grade squamous cell carcinoma arising in a dermoid cyst of ovary. In view of poor prognosis, the patient was kept on follow up.

Discussion

The commonly encountered malignancies in mature cystic teratoma include squamous cell carcinoma followed by adenocarcinoma and carcinoid[5].

Most patients are asymptomatic or present with non specific complaints of abdominal pain, abdominal enlargement, nausea, vomiting, constipation and vaginal bleeding[6]. Examination findings including the abdominal and per-vaginal examination are also non-specific and reveal a pelvic or abdomino-pelvic mass. In our case, the patient presented with abdominal pain and distension.

Hirakawa et al. reported that 3 of 28 MCT patients had previous history of abdominal tumor for 10–32 years, suggesting a long duration of dermoid cyst in the pelvic cavity[7]. This may be due to prolonged exposure to various carcinogens in the pelvic cavity[8] . However no such history was obtained in our case.

The risk factors for malignant transformation in a dermoid cyst include age of the patient, tumor size and image characteristics[3]. This is usually observed in older age in contrast to young age group of dermoid cyst ovary. Chiang et al observed that the median age of patients was 52 years with a mean tumor size of 10.5 cm (range, 1–40 cm)[8]. In the present study, the age at diagnosis of malignant transformation was 52 years.

Along with patient age, the size of the tumor is another clinical characteristic that may raise the suspicion of SCC-MCT, with several reports indicating that these tumors are larger than benign teratomas. Kikkawa et al, in their case series found that mean tumor diameter greater than 9.9 cm was 86% sensitive for squamous cell carcinoma arising in dermoid cyst[9]. In the present case, the tumor size is 9 cm.

Preoperative diagnosis is very difficult due to its rarity and complex components of dermoid cyst. Mostly cases are diagnosed postoperatively as an incidental finding. Serum tumor markers are equivocal in preoperative diagnosis [8]. Chiang et al reported that higher CA125 levels were associated with adverse outcome[8]. Although Hackethal et al. reported no correlation between concentration of tumor markers and FIGO stage, higher levels of markers were predictive of poor survival and prognosis[5]. In our case

serum CA125 was marginally raised. However there are suspicious features of malignant change including: Advanced age of the patient, large size of tumor, solid areas and invasion of adjacent organs/ capsule[3].

Primary SCC of ovary is very rare and usually follows cases of dermoid cyst. This entity carries a worse prognosis than ovarian epithelial cancers of all histologic types[7]. The prognosis is poor especially in advanced stages and in suboptimally cytoreduced patients[10].

Due to its rarity, there is no consensus regarding its management. The optimal management includes debulking surgery and the role of chemotherapy (CT) and radiotherapy (RT) remains unclear[1]. Chen et al. suggested that stage 1A disease, can be treated by conservative surgery[11]. However unilateral salpingo-oophorectomy and surgical staging are proposed for young patients with early stage disease, where preservation of fertility is required[4]. Debulking surgery in the form of TAH-BSO and omentectomy would be the most widely accepted approach, as this allows full staging of the disease[1].

Platinum and Taxane based chemotherapy may be utilized as concurrent treatment strategy. James R Powell in their case series used platinum based chemotherapy and out of six only two patients had partial response[2]. However there was no additional benefit of postoperative RT as demonstrated by Hackenthal *et al*[5].

In summary, preoperative diagnosis of ovarian SCC-MCT is challenging. Clinical, radiologic and intraoperative macroscopic findings are non-specific compared to those of dermoid cyst. A high level of suspicion is required in patients of advanced age with relatively large masses who are thought to have ovarian teratomas. Serum tumor markers might be a useful diagnostic tool to rule out SCC-MCTs. Awareness of this rare entity is required for early diagnosis and better management when dealing with older patients of mature teratoma ovary.

Figure 1: Gross picture showing left ovary solid cystic with friable areas (arrow)

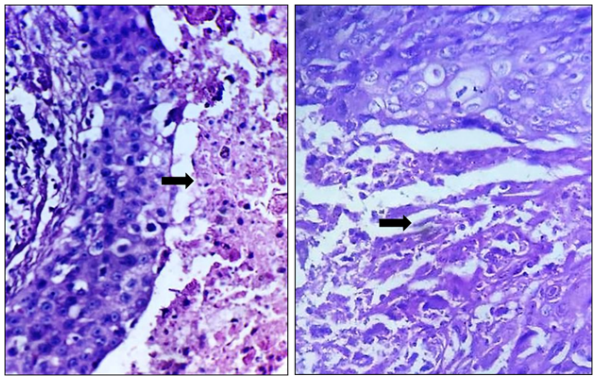

Figure 2: Squamous cell carcinoma in sheets and nests with areas of necrosis (arrow) (H&E x400)

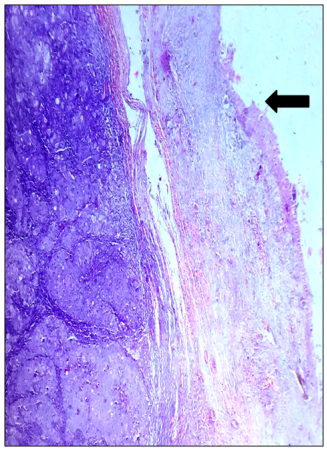

Figure 3: SCC MT from the MCT with nests of tumor cells with hyperchromatic and pleomorphic nuclei; right side shows cyst lined by histiocytes (arrow) (H&E, ×200)

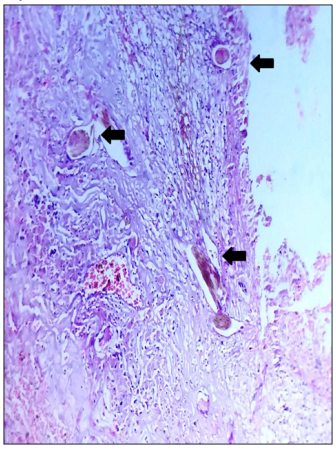

Figure 4: Cyst wall shows embedded hair shaft and follicle (arrow) (H&E x200)

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