CASE REPORT

Squamous cell carcinoma arising in mature cystic teratoma (dermoid cyst) — a rare presentation

Shama Chaudhry, Rubina Hussain

Abstract

Mature cystic teratoma is a common benign adnexal tumour in females. Malignant transformation in a mature cystic teratoma of the ovary is rare, occurring in only 1-2% of cases. The most common malignancy is squamous cell carcinoma, which consists of about 75% of malignant transformations. We describe a 43-year-old female with a 14cm right ovarian mature cystic teratoma with the rare finding of squamous cell carcinoma. The clinical evaluation was done with ultrasound, computed tomography (CT) scan and serologic marker serum CA-125. Exploratory laparotomy was done to remove the mass. Right salpingo-oophorectomy was done. Moderately differentiated invasive squamous cell carcinoma arising in the background of mature cystic teratoma (dermoid cyst) was found. Further, she had total abdominal hysterectomy, left salpingo-oophorectomy and omentectomy, which were disease-free. She is kept on follow-up with ultrasound, CT scan and serum CA-125. Adequately staged patients with disease confined to the ovary have a much better prognosis with 5-year survival rates approaching 95%. Purpose of this case report was to create awareness among physicians while dealing with dermoid cysts of large sizes in older patients.

Keywords: Malignant transformation, Mature cystic teratoma, Squamous cell carcinoma.

Introduction

Mature cystic teratoma (MCT), which is composed of well-differentiated tissues derived from the three germ cell layers (ectoderm, mesoderm, and endoderm), is the most common ovarian tumour. In contrast, squamous cell carcinoma (SCC) of the ovary, which generally has its origin in MCT, is extremely rare.[1] Dermoid cysts (mature cystic teratoma) account for approximately 20% of all ovarian tumours.[2] They occur more commonly in pre-menopausal women and are usually unilateral. Oophorectomy is the operative procedure of choice and is usually curative.[2,3] Purpose of this case presentation is to create awareness among physicians while dealing with dermoid cysts of large sizes in older patients, and also to prepare clinicians and patients mentally for the need of histopathology and further management to decrease mortality by taking timely decision for further surgery, and chemotherapy or radiotherapy.

Clinicians should keep this rare type of tumour in mind when faced with a dermoid cyst, especially in older patients, or in larger than usual cysts.

Case Report

A 43-year-old nulliparous woman presented with pain and enlargement of abdomen for 3 months. She gave no history of weight loss, loss of appetite and no bowel/bladder-related symptoms.

Clinical examinations showed a lump palpable just below the umbilicus. The mass was cystic and non-tender with well-defined margins. No free fluid was detected in the pelvic cavity. A provisional diagnosis of dermoid cyst was made. Ultrasonography showed 14 x 8cm^2 sized ovarian cyst. CT scan (Figure-1) confirmed these findings. Serum CA125 was 9.5 IU/ml.

Exploratory laparotomy was done to remove the mass. Right salpingo-oophorectomy was done. No residual fluid

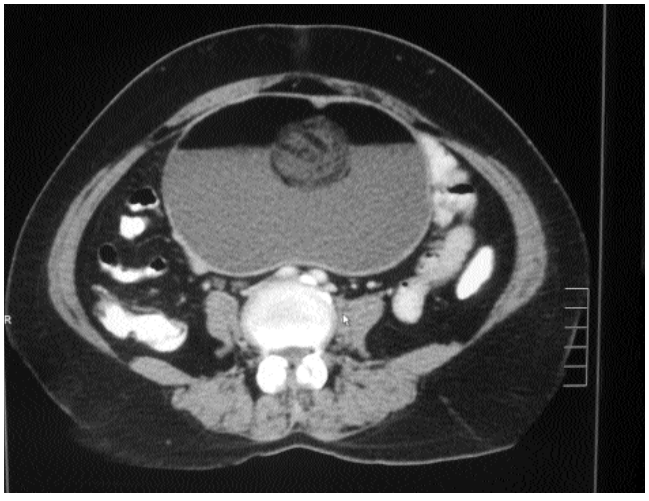

Figure-1: Computed tomography axial view showing well-defined encapsulated cystic lesion with a central solid component and fat-filled fluid level in the centre of abdomen.

Department of Obstetrics and Gynaecology, Ziauddin University, Karachi.
Correspondence: Shama Chaudhry. Email: chshama@yahoo.com

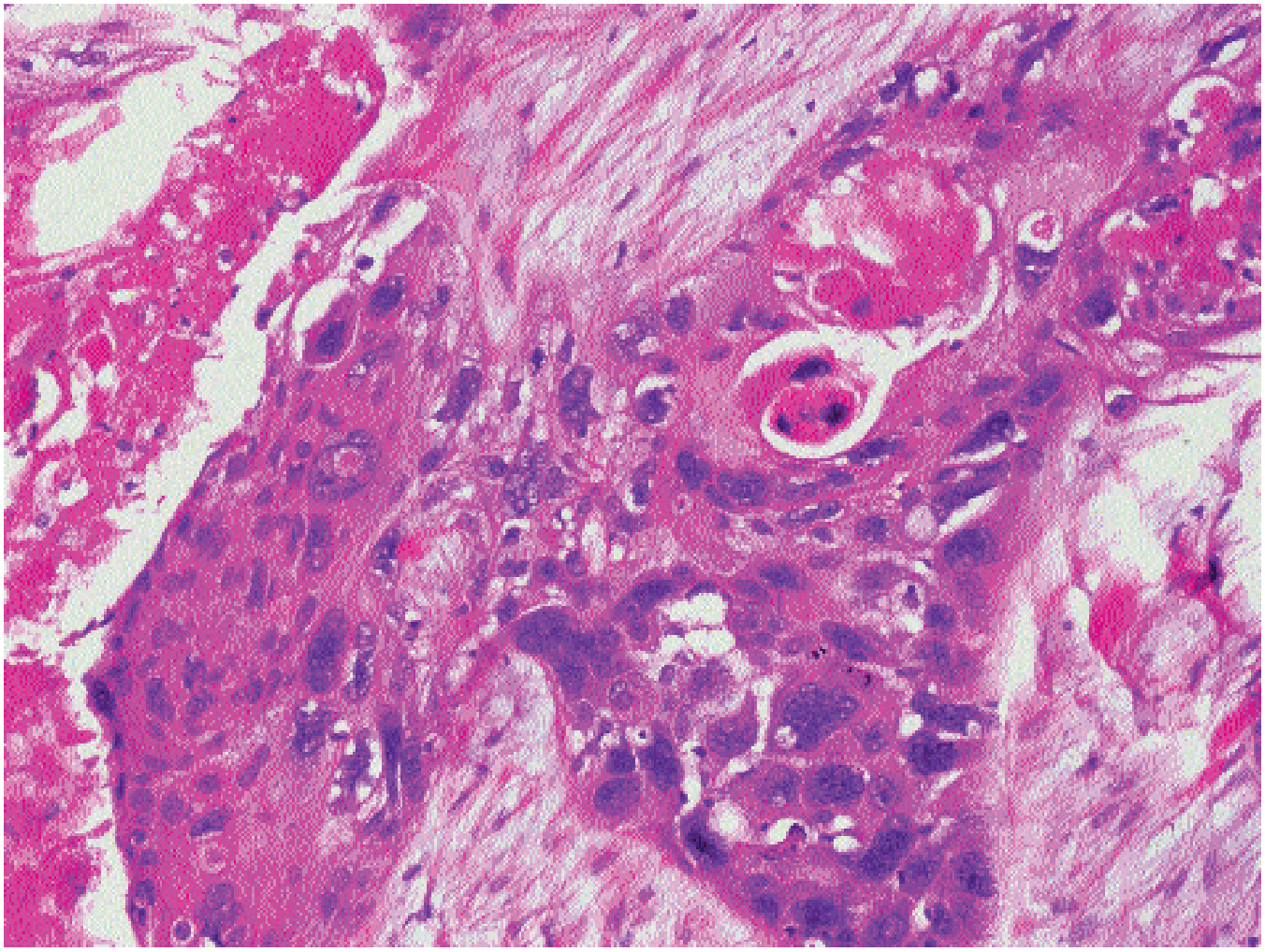

Figure-2: High-powered view showing a nest of malignant squamous cells and keratin pearls.

was seen in the cavity. Left ovary, fallopian tube, and uterus were normal. The ovarian mass measuring 14 x 8 cm^2 in size with a smooth surface was removed along with the fallopian tube. On gross examination, the outer surface was smooth, with attached fallopian tube cyst measured 14 x 12 x 8cm and weighed 983gm. It was a unilocular cyst with small locules seen filled with cheesy material, and contained hair shafts. Focally, an irregular thickened area was identified which was greyish, white necrotic material.

On histopathological examination (Figure-2), sections revealed a fibro-collagenous cyst wall exhibiting a neoplastic lesion. It was composed of strands, cords and nests of squamous cells. These cells showed pleomorphic hyperchromatic nuclei. Foci of keratinisation were noted. Surrounding stroma revealed desmoplasia. Focally, cyst wall was lined by skin with adnexa and showed a cystic area lined by cuboidal ciliated epithelium. Sections from the fallopian tube were tumour-free. Final diagnosis of invasive SCC arising in the background of mature cystic teratoma (dermoid cyst) was made. Histologic grade was moderately differentiated. Further surgery was done and the patient had total abdominal hysterectomy, left salpingo-oophorectomy and omentectomy which were disease-free surgically and histopathologically. It was stage 1A SCC arising in MCT. She is kept on follow-up with ultrasound, CT scan and serum CA-125. The present case carried histological rarity.

Discussion

Malignant transformation in a dermoid cyst of the ovary is a rare complication occurring in only 1-2% of cases, with SCC being the most common type. Pre-operative diagnosis is difficult because of lack of specific symptoms and signs to suggest malignancy.[4] The common symptom

is abdominal pain followed by abdominal or pelvic mass, but the patients may be asymptomatic or have symptoms of abdominal distention or bloated abdomen, as those caused by benign cysts. In some other cases, various symptoms due to invasion of nearby organs are the presenting complaints, such as gastrointestinal symptoms of constipation or diarrhoea, rectal bleeding, or urinary frequency. Other non-specific signs of wasting disease, such as weight loss or cachexia may be found in advanced cases.

Pre-operative diagnosis of an MCT of the ovary is relatively easy due to the radiological detection of bony tissues, including teeth, bones and cartilages. However, pre-operative diagnosis of malignant transformation is very difficult clinically, because this tumour cannot be readily differentiated from an uncomplicated MCT or other ovarian tumours. SCC arising in an MCT has historically been observed in relatively older patients, particularly after menopause; though it has sometimes been reported in young patients around 30 years.[5] Although germ cell tumours generally occur in younger patients, SCC arising from MCT occurs in patients who are older than those who develop other malignant germ cell tumours.[1] Tumour size has also been noted to predict malignancy. Although MCT presents in a wide range of sizes, larger tumours correlate with an increased risk of malignant transformation. In our case, the tumour diameter was around 14cm, which is larger than a typical benign cyst. A study reported that a tumour diameter of larger than 9.9cm was 86% sensitive for malignancy.[1] At the time of presentation, the most frequent symptom associated with malignant transformation is lower abdominal or pelvic pain and increasing abdominal girth.[6] Old age, large tumour size, and solid portion in MCT seem to predict the malignant transformation of MCT. The prognosis of SCC is much worse than that of other epithelial ovarian cancers.[1] SCC arising in MCT has a poor prognosis when the disease has spread beyond the ovary. Adequately staged patients with disease confined to the ovary have a much better prognosis with 5-year survival rates approaching 95%.[7]

The main therapeutic approach to an ovarian MCT with malignant transformation has been surgical. Conservative unilateral oophorectomy without further post-operative treatment may be justified for early stage IA disease, especially for nulliparous and young patients who desire future fertility; however, in the post-menopausal women, total removal of the genital organs would seem to be the procedure of choice.[8] Post-operative treatments in the literature included single-agent or combination chemotherapy, radiotherapy, or a combination of these modalities. Results of these treatment regimens were variable and have not been systemically evaluated in an adequate number. Therefore, the optimal adjuvant therapy for SCC arising from an MCT has not been yet established.[3]

Conclusion

Clinicians should keep this rare type of tumour in mind when faced with a dermoid cyst, especially in older patients, or in patients with larger than usual cysts.

References

1. Kikkawa F, Nawa A, Tamakoshi K, Ishikawa H, Kuzuya K, Suganuma N, et al. Diagnosis of squamous cell carcinoma arising from mature cystic teratoma of ovary. Cancer 1998; 82: 2249-55.
2. Powell JL, Stinson JA, Connor GP, Shiro BS, Mattison M. Squamous cell carcinoma arising in a dermoid cyst of the ovary. Gynecol Oncol 2003; 89: 526-8.
3. Tangjitgamol S, Manusirivithaya S, Sheanakul C, Leelahakorn S, Thawaramara T, Jesadapatarakul S. Squamous cell carcinoma arising from dermoid cyst: Case reports and review of literature. Int J Gynecol Cancer 2003; 13: 558-63.
4. Hurwitz JL, Fenton A, McCluggage WG, McKenna S. Squamous cell carcinoma arising in a dermoid cyst of the ovary: a case series. BJOG 2007; 114: 1283-7.
5. Mayer C, Miller DM, Ehlen TG. Peritoneal implantation of squamous cell carcinoma following rupture of a dermoid cyst during laparoscopic removal. Gynecol Oncol 2002; 84: 180-3.
6. Lim SC, Choi SJ, Suh CH. A case of small cell carcinoma arising in mature cystic teratoma of ovary. Pathol Int 1998; 48: 834-9.
7. Shariat-Torbaghan S, Emami-Aleagha M, Sedighi S, Azadbakht F, Keshvar A, Hajarizadeh B, et al. Squamous cell carcinoma arising in an ovarian mature cystic teratoma: a case report. Arch Iran Med 2009; 12: 186-9.
8. Tseng CJ, Chou HH, Huang KG, Chang TC, Liang CC, Lai CH, et al. Squamous cell carcinoma arising in mature cystic teratoma of the ovary. Gynecol Oncol 1996; 63: 364-70.