Case Report

Intestinal Adenocarcinoma Arising from a Mature Cystic Teratoma

King Man Wan,1 Forough Foroughi,2 Rajni Bansal,3 and Martin K. Oehler1,4

1Department of Gynaecological Oncology, Royal Adelaide Hospital, Adelaide, South Australia, Australia
2Department of Anatomical Pathology, Royal Darwin Hospital, Darwin, Northern Territory, Australia
3Department of Obstetrics and Gynaecology, Alice Springs Hospital, Alice Springs, Northern Territory, Australia
4Discipline of Obstetrics and Gynaecology, Adelaide Medical School, University of Adelaide, Adelaide, South Australia, Australia

Correspondence should be addressed to Martin K. Oehler; oehler.mk@gmail.com

Received 14 July 2019; Accepted 6 September 2019; Published 18 November 2019

Academic Editor: Showket Hussain

Copyright © 2019 King Man Wan et al. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Mature cystic teratomas are the most common ovarian germ cell tumour and account for 10–20% of all ovarian neoplasms. Malignant transformation is rare and reported to occur in approximately 0.17–0.8% of cases [1, 2]. The most common malignancy are squamous cell carcinomas. Here we present the case of an intestinal adenocarcinoma which is an exceedingly rare malignant entity arising within a mature cystic teratoma. Clinical presentation, imaging and histopathological diagnosis are discussed and previously presented cases in the literature reviewed.

1. Introduction

Mature cystic teratoma (MCT) of the ovary accounts for 10–20% of all ovarian neoplasms and is the most common ovarian germ cell tumour. Malignant transformation is rare and reported to occur in approximately 0.17–0.8% of cases [1, 2]. The most common malignancy are squamous cell carcinomas but basal cell carcinomas, sebaceous tumours, malignant melanomas, adenocarcinomas, sarcomas, and neuroectodermal tumours have also been reported [3].

2. Case Presentation

A 58-year-old woman living in a remote region was referred to the local general gynaecological service for investigation of an episode of light postmenopausal bleeding. A pelvic ultrasound demonstrated a right sided complex adnexal mass measuring 101 × 70 × 89 mm and a borderline endometrial thickness of 5.1 mm. The mass had a well circumscribed outer capsule with no evidence of increased internal vascularity and there was not ascites. A CT of the abdomen and pelvis demonstrated the complex mass containing internal calcification, fluid, fat and soft tissue (Figure 1). There was no evidence of peritoneal or omental metastasis. The tumour markers showed an elevated CA 19-9 of 58 and normal CA 125 (14), CEA (2), AFP (4.1), HCG (<1) and LDH (220). The complete blood count as well as renal and liver functions were normal.

The patient’s medical history was unremarkable and included one normal vaginal delivery. She had gone through menopause at age 48 and had not used any HRT. In her family history, she only had her sister with an early-stage endometrial adenocarcinoma.

The patient was referred to a tertiary gynaecological oncology service and after discussion at a multi-disciplinary team meeting with radiology review, the tentative diagnosis of mature teratoma with low risk for malignancy was made.

A hysterectomy, dilatation and curettage of the uterus were performed and showed atrophic endometrium. The patient then underwent a total abdominal hysterectomy and bilateral salpingo-oophorectomy. Intraoperatively, she was found to have a mobile 10 cm left ovarian mass with no surface excrescences. The lesion was removed intact. The right adnexa, uterus and peritoneum were normal. Peritoneal washings were obtained. The operation was uncomplicated, and the patient was discharged from hospital on day 3 after the procedure.
Macroscopically, the left ovarian cyst measured 100 × 80 × 70 mm in diameter and had a maximum wall thickness of 2 mm. It contained yellow cheesy material, teeth and hair. The uterus, cervix and right ovary were unremarkable and histologically normal.

Histology of the ovarian cyst showed mature cystic teratoma, predominantly comprised of skin and adnexal elements with large areas of foreign-body type granulomatous response to hair. One section of the cyst was lined by dysplastic columnar type epithelium in continuity with squamous epithelium. Within this area, there were atypical irregular glands infiltrating the underlying stroma. The atypical glands were lined by pleomorphic cells with hyperchromatic enlarged nuclei and luminal dirty necrosis (Figures 2(a)–2(d)).

Immunohistochemical staining showed the tumour cells were strongly positive for CK20, CDX2, focally positive for CK7 and negative for CD30, PAX-8, Vimentin, ER, PR, and CD10 (Figures 3(a)–3(c)). MUC-2 staining was also performed and
showed cytoplasmic positivity in tumoural cells consistent with intestinal differentiation (Figure 3(d)). Perineural invasion was identified, but no lympho-vascular invasion or surface involvement. No immature elements were found. The final diagnosis was intestinal-type moderately differentiated adenocarcinoma arising within a mature cystic teratoma.

The case was re-discussed at the multidisciplinary gynaecological oncology meeting and the disease staged as FIGO Stage 1A ovarian intestinal adenocarcinoma arising within a mature cystic teratoma. She was recommended to have adjuvant platinum-based chemotherapy to decrease risk of recurrence. However, after medical oncology review, the patient elected for observation only. She is alive and well after 12 months of follow up.

3. Discussion

MCTs or dermoid cysts are the most common ovarian germ cell tumour. They arise from totipotent cells in the ovary which develop into fully differentiated ectodermal, mesodermal, and endodermal tissue. Parthenogenetic activation of oocytes (embryonic development without a male gamete) is the most widely accepted theory for the origin of MCTs, primarily because of presence of 46, XX karyotype in almost all mature teratomas [4].

Intestinal adenocarcinomas arising within cystic teratomas are exceedingly rare and this is only the 12th reported case in the literature (Table 1). Intestinal adenocarcinomas are suspected to arise from the endodermal cell line with prevailed derivation of the lower gastrointestinal tract structure, thereby demonstrating characteristics of intestinal differentiation in immunohistochemistry, with CK20 and CDX2 positivity [5, 6]. Focal positivity for CK7 in our case was misleading initially as it suggests a primary ovarian mucinous tumour. However, subsequent positive staining with MUC2 confirmed the diagnosis.

The mechanisms of malignant transformation in ovarian MCTs are uncertain. MCTs are thought to result from replication errors during meiosis and they may represent primary oocytes that have escaped from meiotic arrest. However, it is unclear how subsequent malignant transformation occurs. A systematic genomic evaluation of ovarian SCC arising in MCT showed similarities to other non-HPV SCC, but with distinct features, including bi-allelic TP53 mutations [7]. Further research will be required to address the question of MCT cell of origin and to understand what causes transformation of the MCT into malignancies.

It has been suggested that prolonged exposure of MCTs to carcinogens in the pelvic cavity might promote malignant transformation, as MCTs are usually detected 15–20 years earlier than malignant transformations [8]. If this might warrant a more pro-active removal of MCTs to avoid long term malignant transformation is unknown.

Predictive factors for malignant transformation of MCTs include old age, large tumour size, raised CA125, postmenopausal status and presence of solid components [6]. However, 80% of malignant transformations have been reported in women of reproductive age [7]. Ultrasound imaging may show branching isoechoic components and magnetic resonance imaging
especially fat-suppression images, may increase preoperative suspicion for malignant transformation [9, 10]. Tumour markers are inconsistently reported but elevated CEA and CA19.9 may indicate malignant transformation. Elevated serum squamous cell carcinoma (SCC) antigen levels can help differentiating between benign and MCTs with malignant transformation, but cell carcinoma (SCC) antigen levels can help differentiating between colorectal adenocarcinomas and extra-intestinal gastrointestinal adenocarcinomas [12]. However, histopathology cannot differentiate between primary intestinal adenocarcinoma and metastatic carcinomas [13]. CDX2, CK20 and CK7 helps to differentiate between colorectal adenocarcinomas and extra-intestinal gastrointestinal adenocarcinomas [12]. However, histopathology cannot differentiate between primary intestinal adenocarcinoma and metastatic carcinomas [13].

**4. Conclusions**

Intestinal-type adenocarcinoma arising in MCTs are very rare malignancies and preoperative diagnosis is difficult. Histopathological diagnosis is aided by the use of the
immunohistochemical markers, CK 7, CK20 and MUC2. Most cases are diagnosed at Stage 1 and tend to have a better prognosis than other forms of malignant transformation.

**Conflicts of Interest**

The authors declare that they have no conflicts of interest.

**References**


