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CASE REPORT

Squamous cell carcinoma of the ovary arising in a mature cystic teratoma in a 28-year-old female: a case report and review of literature

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Malignant transformation of mature cystic teratoma is a rare phenomenon occurring in approximately 0.17–2% of cases. It has frequently been noted in peri- and postmenopausal women, but it is rarely seen in younger women. There are no distinctive clinical features. However, patients present with nonspecific signs and symptoms such as abdominal pain, distension and abdominal mass. Hysterectomy, bilateral salpingo-oophorectomy and comprehensive surgical staging are the cornerstone of surgical management. In advanced disease, optimal cytoreduction is associated with increased overall survival. While there is no consensus as to the optimal adjuvant therapy, chemotherapy with an alkylating drug has been noted to have the greatest impact on survival. Advanced-stage disease has a poor prognosis, worse than that of epithelial ovarian cancers. We report on a case of a 28-year-old female who presented an abdominal swelling of 10 months, having undergone two previous laparotomies. Histopathology of the left ovary showed features suggestive of invasive squamous cell carcinoma and of the right ovary, it showed features suggestive of dermoid cyst. No tumour deposits were noted on the omentum. The left ovary ruptured during the procedure upstaging the tumour. An attempt at cytoreduction was done at our facility. However, tumour was noted to be unresectable. She received two cycles of carboplatin and paclitaxel but died of the disease three months later.

Keywords: squamous cell carcinoma, mature cystic teratoma, malignant transformation, ovarian neoplasm

Background

Malignant transformation of mature cystic teratoma (MCT) is a rare phenomenon occurring in approximately 0.17–2% of cases.[1] Although any component of the MCT may undergo transformation, 80% of malignant transformations are to squamous cell carcinoma (SCC) arising from the ectoderm.[2,3] This transformation is frequently noted in postmenopausal women.[2,4,5]

Patients present with nonspecific signs and symptoms such as abdominal pain, distension and abdominal mass.[5] Tumour markers such as cancer antigen 125 (CA 125), squamous cell carcinoma (SCC) antigen, alpha-fetoprotein (AFP), human chorionic gonadotropin (HCG) and lactate dehydrogenase (LDH) are often normal.[2] Preoperative radiological diagnosis is difficult. However, magnetic resonance imaging (MRI) may also be helpful. Kido et al.[6] correlated malignant transformation in MCT with the presence of solid components with contrast enhancement, transmural or transseptal extension, evidence of adherence to surrounding structures, necrosis, and haemorrhage. Malignant transformation of an ovarian cyst, therefore, is difficult to predict. Hence, most cases are diagnosed postoperatively. Cytoreductive surgery and proper staging is usually recommended, and in younger women with disease confined to the ovary, fertility sparing surgery may be considered. Owing to the relative rarity of this disease, there is no consensus on the adjuvant management. However, chemotherapy and radiotherapy have been used following surgery.[3,4] While prognosis depends upon the stage at presentation, disease confined to the ovary has a better prognosis.[5]

Case presentation

We present a case of SCC arising from MCT in a 28-yr-old Para 2+0, who presented with an abdominal swelling and distension for 10 months, having undergone two previous laparotomies by a general gynaecologist. The initial surgery was done four months prior to presentation for bilateral ovarian masses, and bilateral salpingo-oopherectomy and infracolic omentectomy were done. Intraoperatively the patient was noted to have bilateral ovarian masses. The left mass was solid with cystic areas, approximately 18 cm x 15 cm x 11 cm with an intact capsule, which ruptured during the procedure. The right mass was cystic, measuring 20 cm x 13 cm x 8 cm, with an intact capsule. The uterus and the rest of the abdomen appeared grossly normal. Histopathology of the left ovary showed features of moderately differentiated SCC, while the right ovary showed features of dermoid cyst with areas of severe squamous dysplasia. Both fallopian tubes were unremarkable. The omentum showed no evidence of malignancy. The patient underwent a second surgery 14 weeks later due to abdominal distension, and extensive tumour with seedings were seen intraoperatively all over the abdominal cavity.

At presentation, the patient was wasted and pale, with an Eastern Cooperative Oncology Group status of 2 with an abdominal mass of 20 cm x 10 cm and a normal pelvic exam. Investigations

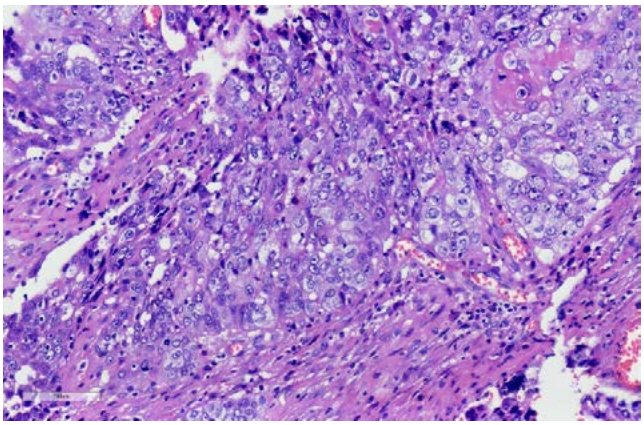

Figure 1: Tumour from the mesenteric nodules show pleomorphic epithelial cells forming infiltrating nest and cords (H & E x 20)

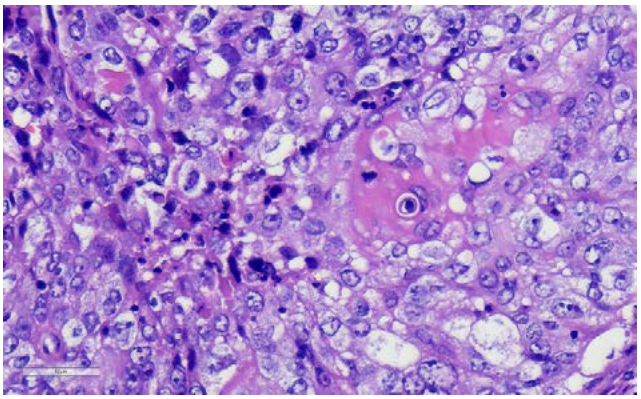

Figure 2: Neoplastic cells exhibit prominent intercellular bridges and cytoplasmic keratinisation – mitotic activity is brisk

indicated a white blood cell count of 30 000, with a neutrophilia of 25 000, haemoglobin level of 7.8g/dl, platelet count of 549, LDH levels were elevated at 267U/L, CA-125/AFP/ carcinoembryonic antigen/carbohydrate antigen 19-9 were within normal range, and the pap smear was unremarkable. A computerised tomography (CT) scan of the abdomen/pelvis showed multiple irregular thick-walled abdominal pelvic cystic collections with areas of the small bowel wall thickening and swelling of the uterus, suggestive of infective process with early mesenteric abscess. A third laparotomy was done. Intraoperatively, the left anterior abdominal wall septations with necrotic mass, dense adhesions, multiple matted mesenteric nodules and tumour were seen on the transverse colon and rectum. The uterus, liver, spleen and paracolic gutters appeared grossly normal. Adhesiolysis was done, the abdominal wall abscess was drained and multiple biopsies were taken. Histopathology of the sigmoid, mesenteric and small gut nodule showed features consistent with SCC.

Postoperatively, the patient's condition improved markedly and she received two cycles of the carbotaxol. Three months later, however, she died of the disease.

Discussion

Primary SCC is a rare entity that usually arises from the malignant transformation of MCT, and rarely from endometrioma and Brenner tumour.[7] The most common pathological presentation of ovarian SCC is from transformation of MCT which may contain squamous cells. A histopathologic study revealed that SCC may arise not only from the epidermis, but also from squamous metaplastic epithelium of respiratory glands.[5] The pure variety is very rare and arises from the metaplasia of the surface epithelium of the ovary.[3,8] SCC is mostly observed in postmenopausal women in contrast to MCT which usually occurs in younger women, though some SCCs have been reported in women as young as 19 years of age.[3,9] Patients are usually asymptomatic, although some may present with abdominal pain and an abdominopelvic mass.[9] Tumour size may range from 10–40 cm.[3,10,11]

Management of SCC of the ovary is similar to that of other epithelial carcinoma. Hysterectomy, bilateral salpingo-oophorectomy, comprehensive surgical staging with peritoneal washing cytology, omentectomy, peritoneal biopsy and pelvic plus para-aortic lymphadenectomy, form the cornerstone of surgical management.[12,13] In advanced disease, optimal cytoreduction is associated with increased overall survival compared to suboptimal cytoreduction.[13] Kashimura et al.[5] reported that patients who did not undergo complete surgical resection but received intraperitoneal chemotherapy and radiotherapy, died within one year. For patients who were not optimally staged at initial surgery, a second laparotomy should be performed as complete excision of the tumour improves prognosis.[13] Stage IA disease can be treated with conservative surgery for young women who desire fertility, as patients who undergo conservative surgery for stage IA disease after surgical staging have the same survival rate as those who receive more extensive surgery.[13,14] Early disease, particularly that confined to the ovaries, has been noted to have a good prognosis, with 2- and 5-year overall survival of 100% and 75%, respectively.[13,14]

The benefits of adjuvant treatment with chemotherapy and radiotherapy are unclear. The most commonly used chemotherapy regimen is carboplatin or cisplatin with paclitaxel.[2,13,14] Chemotherapy with an alkylating agent is associated with higher survival rates in tumour stages greater than 1A, while no improvement was noted using radiotherapy.[2] Similarly, another review reported improved survival rates for stages II/III/IV with platinum-based chemotherapy, while there is no benefit with radiotherapy.[9] Chen et al.[13] reported no benefit with chemotherapy in stage I/II and significant improvement in survival rates in stage III/IV, while no benefit in any of the stages with radiotherapy.

The prognosis of patients with malignant transformation of MCT is worse than that of patients with common epithelial ovarian cancer.[12] The prognosis depends heavily on the stage of the disease and optimal debulking.[13] Other factors affecting diagnosis are tumour grade, tumour dissemination, cyst-wall invasion, rupture, ascites, adhesion, growth pattern and vascular invasion.[2,9,13] A systematic review reported an overall 5-year survival rate for all stages to be 48.4%, and for stage I, II, III and IV to be 75.7%, 33.8%, 20.6% and 0%, respectively.[13] Hackethal et al.[2] reported striking differences between stage I disease and other stages with only minor prognostic differences between stages II, III and IV.

Conclusion

SCCs arising in MCT are commonly large ovarian tumours that occur in postmenopausal women. There is a lack of consensus regarding adjuvant treatment. Complete staging with hysterectomy plus bilateral salpingo-oophorectomy and surgical cytoreduction for anyone with extra-ovarian disease, followed by adjuvant chemotherapy with an alkylating drug, have the greatest impact on survival. Advanced-stage disease has a poor prognosis, worse than that of epithelial ovarian cancers.

Conflict of interest

The authors declare no conflict of interest.

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