CASE REPORT

Ovarian squamous cell carcinoma arising from mature cystic teratoma

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Abstract

Preoperative diagnosis of malignant transformation of an ovarian mature cystic teratoma to squamous cell carcinoma is difficult due to nonspecific tumor markers and imaging findings. This is an interesting case presentation that has prior imaging that demonstrates imaging characteristics of the transformation of a mature cystic teratoma to squamous cell carcinoma.

Keywords: Ovarian squamous cell carcinoma; malignant transformation; ovarian mature cystic teratoma.

Clinical presentation

A 68-year-old woman presented with a 9-kg weight loss, low-grade fever, fatigue, abdominal cramping and pain. Her past medical history was notable for Crohn disease status post right hemicolectomy 14 years ago, left breast cancer (ductal carcinoma in situ) status post lumpectomy and radiation 5 years ago, and excision of melanoma in the left arm 3 years ago. Physical examination was unremarkable with the exception of a firm lower abdominal mass. Liver function and renal function values were normal and complete blood count demonstrated mildly elevated white count at 14 × 10^3 cells/ml, normal hemoglobin (12.4 g/dl) and hematocrit (37.4%). No tumor markers were performed at this time. Computed tomography (CT) of the abdomen and pelvis showed a left pelvic mass measuring 13 × 8 × 12 cm containing fat, soft tissue and calcification (Fig. 1). The left ovary was not visualized separately. The right ovary appeared normal. The mass displaced the uterus to the right, which contained calcified fibroids. The mass was adherent to the sigmoid colon (Fig. 2), and obstructed the distal left ureter resulting in mild left hydroureteronephrosis. There was no ascites or pelvic side wall extension.

Prior imaging studies included a CT of the abdomen/pelvis 9 years previously that had demonstrated a 9 × 6 cm fat-containing left adnexal mass (Fig. 3). Compared with the previous image, the mass had significantly enlarged in size and demonstrated a new, large, heterogeneous soft tissue component within the previously fat-containing mass. The increase in size and development of a soft tissue component, along with new symptoms, was highly concerning for malignant transformation. The patient was advised to proceed with an exploratory laparotomy and surgical removal of the mass.

At surgery, the left ovary was found to be replaced with a 13-cm solid mass with a 4-cm tumor plaque densely adherent to the left pelvic side wall and the rectosigmoid colon. Intraoperative frozen section of the left ovarian mass revealed squamous cell carcinoma. The patient underwent total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy, bilateral pelvic lymph node dissection, excision of tumor from the surface of the sigmoid and left pelvic side wall. Peritoneal washings for cytology were negative. Gross pathology showed a multicystic mass, measuring 15.0 × 11.0 × 7.5 cm, with a central cystic/necrotic mass measuring...
8.0 x 7.0 x 7.0 cm containing a collection of hair and liquefied/necrotic fat. The cyst walls were thick and fleshy measuring up to 2.5 cm. Histopathology showed that the majority of the left ovary was replaced by malignant epithelium (Fig. 4) with little benign mature squamous epithelium remaining consistent with squamous cell carcinoma, moderate to poorly differentiated, developing within mature cystic teratoma. There was no lymphovascular invasion or perineural invasion. The cervix, endometrium, myometrium were normal. Left external iliac lymph nodes, left obturator lymph node tissue, right ovary, right external iliac lymph node tissue, right obturator lymph node and omentum were all negative for tumor.

Mature cystic teratoma (MCT) is a common ovarian neoplasm\(^1,2\). Malignant transformation of an MCT is an uncommon event occurring in less than 2% of all mature dermoid cysts\(^3\). The most common secondary malignancy to arise from a MCT is an invasive squamous cell carcinoma. Other malignancies include adenocarcinoma, sarcoma, carcinoid, thyroid carcinoma and melanoma\(^4\). Non-teratomatous squamous cell carcinoma of the ovary has been found to originate from endometriosis, epidermoid cysts or from the surface epithelium\(^5\).

Age and size are predictors of malignant transformation in benign teratoma. Malignant transformation tends to occur more commonly in older women with an average mean age at diagnosis of 55 years\(^3\). Malignant transformed mature cystic teratomas have a larger size (mean size of 15 cm) compared with MCTs (mean size of 6–9 cm)\(^3\). Symptoms at presentation are variable in both diagnoses with some patients presenting with acute abdominal pain and others with constitutional symptoms such as fatigue, urinary symptoms and anorexia/weight loss\(^6\).

Although tumor markers may be raised in patients with squamous cell carcinoma arising from MCT, it is difficult to use tumor markers to distinguish between MCT and
squamous cell carcinoma arising from an MCT since tumor markers can also be elevated with MCT\textsuperscript{[7]}\textsuperscript{[7]}. For example, squamous cell antigen (SCC) levels were found to be significantly higher in patients with squamous cell carcinoma arising from MCT than with MCT alone\textsuperscript{[8]}\textsuperscript{[8]}. However, mean levels in squamous cell carcinomas are lower than in patients with adenocarcinomas, and cannot predict the diagnosis preoperatively\textsuperscript{[8]}\textsuperscript{[8]}. CA19-9 is another tumor marker found to be significantly higher in patients with squamous cell carcinoma arising from MCT than with MCT alone, however it is also a difficult marker to use in preoperative screening since the mean levels of CA 19-9 are found to be elevated in patients with MCT alone\textsuperscript{[8]}\textsuperscript{[8]}.

Radiologically, mature teratomas may demonstrate a broad spectrum of findings ranging from a purely cystic mass, a fat-containing mass or a heterogeneous soft tissue mass. Classic diagnostic findings for MCT include fat attenuation within a cyst that possibly also contains calcification\textsuperscript{[9]}\textsuperscript{[9]}. Imaging features concerning for malignant transformation include thick walls, enhancing solid components or papillary projections within the cyst, peritoneal deposits or lymphadenopathy. In our case the availability of prior CT imaging demonstrating a fat-containing adnexal mass with areas of calcification and a subsequent CT that demonstrated interval enlargement with a new, enhancing soft tissue component was concerning for malignant transformation of the MCT.

MCT can be managed expectantly in premenopausal women with ovarian dermoid cysts less than 6 cm in diameter, especially if pregnancy is desired. The mean growth rate of dermoid cysts in premenopausal women is 1.8 mm/year\textsuperscript{[10]}\textsuperscript{[10]}. However, if tumor markers are abnormal and worrisome imaging findings are noted, a surgical approach is advised\textsuperscript{[7]}\textsuperscript{[7]}. Ovarian torsion and rupture are other complications of MCT to consider in addition to malignant degeneration\textsuperscript{[9]}\textsuperscript{[9]}. Masses such as MCT that can predispose the ovary to torsion can also mimic torsion\textsuperscript{[11]}\textsuperscript{[11]}. The most common imaging finding of ovarian torsion is ovarian enlargement and cross-sectional imaging may show lack of enhancement\textsuperscript{[11]}\textsuperscript{[11]}. Enhancing solid components and/or papillary projections would be less characteristic of ovarian torsion and rather of malignant degeneration.

Given the rarity of squamous cell carcinoma of the ovary, there is no clear standard of care and often patients are treated similar to patients with epithelial ovarian cancer\textsuperscript{[7]}\textsuperscript{[7]}. Review of multiple case series recommends surgery including total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy and lymph node dissection\textsuperscript{[6,7]}\textsuperscript{[6,7]}. Standard treatment includes chemotherapy with a platinum-based agent. The role of radiotherapy remains undefined\textsuperscript{[6,7]}\textsuperscript{[6,7]}. The stage of disease is the best predictor of survival with overall better outcome for patients with limited disease with disease-free survival for up to 5 years after diagnosis\textsuperscript{[6]}\textsuperscript{[6]}. Our patient was considered a high risk for recurrence given tumor deposits on the pelvic side wall and colon and was treated postoperatively with 6 cycles of a platinum and paclitaxel. Although radiation was considered, the decision was made against treatment given presumed possible toxicity associated with inflammatory bowel disease. She remains disease free on surveillance imaging at 18 months since resection.

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