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clinical practice

A rare case report of squamous-cell carcinoma arising from mature cystic teratoma of ovary

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SUMMARY: **A rare case report of squamous-cell carcinoma arising from mature cystic teratoma of ovary.**

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The most frequent ovarian germ cell tumors are mature cystic teratomas (MCTs), composing 10-25% of all ovarian neoplasms. MCTs have the potential of undergoing malignant transformation, typically in postmenopausal women, with a frequency of 0.17-3%, with squamous cell carcinoma being the most common malignant tumor arising from MCT.

We present the rare clinical entity of a squamous cell carcinoma arising from a mature cystic teratoma in a 56-year-old premenopausal woman as well as diagnostic and therapeutic route followed.

KEY WORDS: Ovary - Teratoma - Carcinoma.

Introduction

The most frequent ovarian germ cell tumors are mature cystic teratomas (MCT$_s$), composing 10-25% of all ovarian neoplasms and 5% of ovarian cancers (1, 2). It is believed that they arise from postmeiotic germ cells (3), consisting of all three germ-cell layers (ectoderm, mesoderm, and endoderm) (2).

MCTs grow in the fifth to sixth decade of a woman's life (4); nevertheless they are also very common in women of childbearing age, found in both ovaries in 10-17% of patients (5, 6). Their clinical presentation seems to be similar of all ovarian tumors as they cause abdominal pain, constipation, bleeding, weight loss, urinary frequency and fever (1, 3).

The potential of undergoing malignant transformation (one or more of the three different mature elements of MCTs) is present, typically in postmenopausal women, with a frequency of 0.17-3% (7-10). Most of MCTs are detected 10-15 years before secondary malignant transformation possibly as a result of exposure in different pelvic carcinogens which trigger malignant changes in mature tissue (11). Due to the high density of ectoderm in these tumors, not surprisingly the most common malignant tumor arising from them is squamous cell carcinoma (SCC) (2, 10); while various adenocarcinomas, carcinoid tumors, melanomas and various soft tissue sarcomas have also been reported (2, 4, 12).

Case report

A 56-year-old, gravida 2, para 2, premenopausal, Caucasian woman presented to our hospital with abdominal cramping and pain. Her past medical history was free and physical examination was unremarkable with the exception of a firm lower abdominal mass.

Transabdominal and transvaginal ultrasound showed a 6.27 x 10 cm pelvic mass containing fat, soft tissue and calcification (Fig. 1). Computed tomography (CT) of the abdomen and pelvis showed a right pelvic mass measuring 10x6x4 cm. The left ovary appeared normal. Neither ascites nor pelvic nodes were found.

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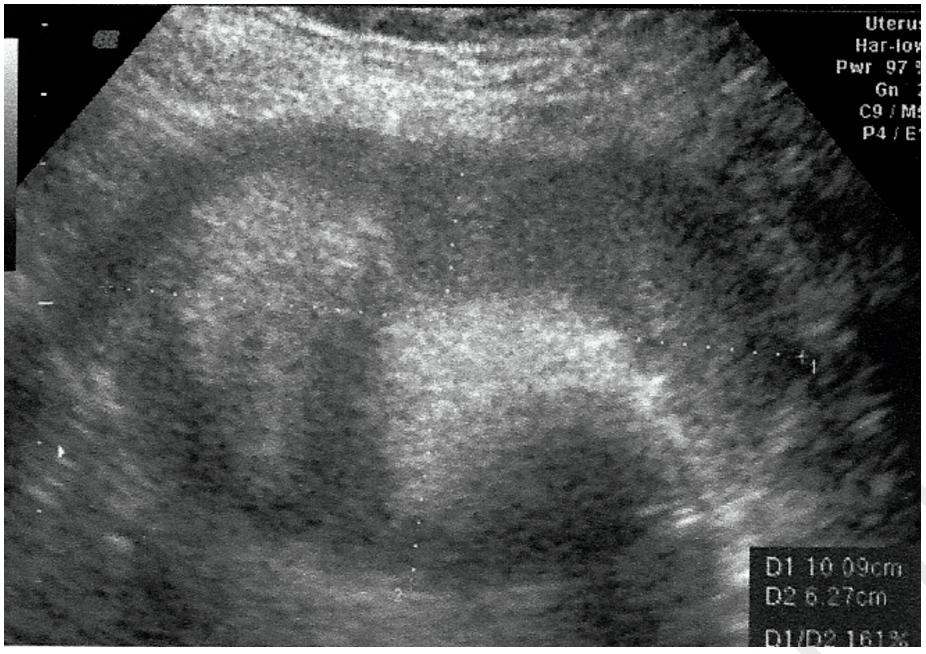

Fig. 1 - Transabdominal ultrasound of pelvic mass.

Complete blood count demonstrated normal hemoglobin (12.5 g/dL) and hematocrit (38%). Liver function and renal function values were normal. Tumor markers (CA-125, CA 19-9, alpha fetoprotein {AFP}, carcinoembryonic antigen {CEA}) were normal too.

The patient underwent exploratory laparotomy. The specimen of "right ovarian mass" consisted of an intact cystic structure, had a smooth external surface, measured 7.5×8.5×10.5 cm in size and weighed 360 grams. It was filled with white greasy sebaceous material intermixed with hair (Fig. 2). Microscopically, keratinising squamous epithelium with underlying skin adnexa, respiratory type epithelium, cartilage and neural tissue were identified. Focally, the cyst lining consisted of non-ke-

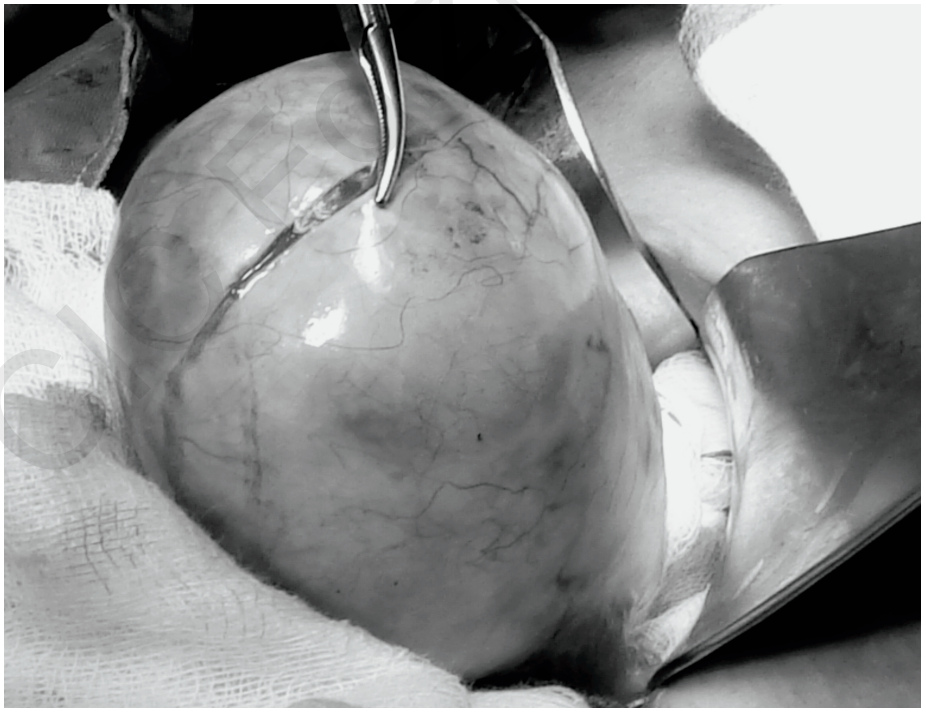

Fig. 2 - Gross appearance of mature cystic teratoma with squamous cell carcinoma. The cyst is filled with cheese-like material and hair.

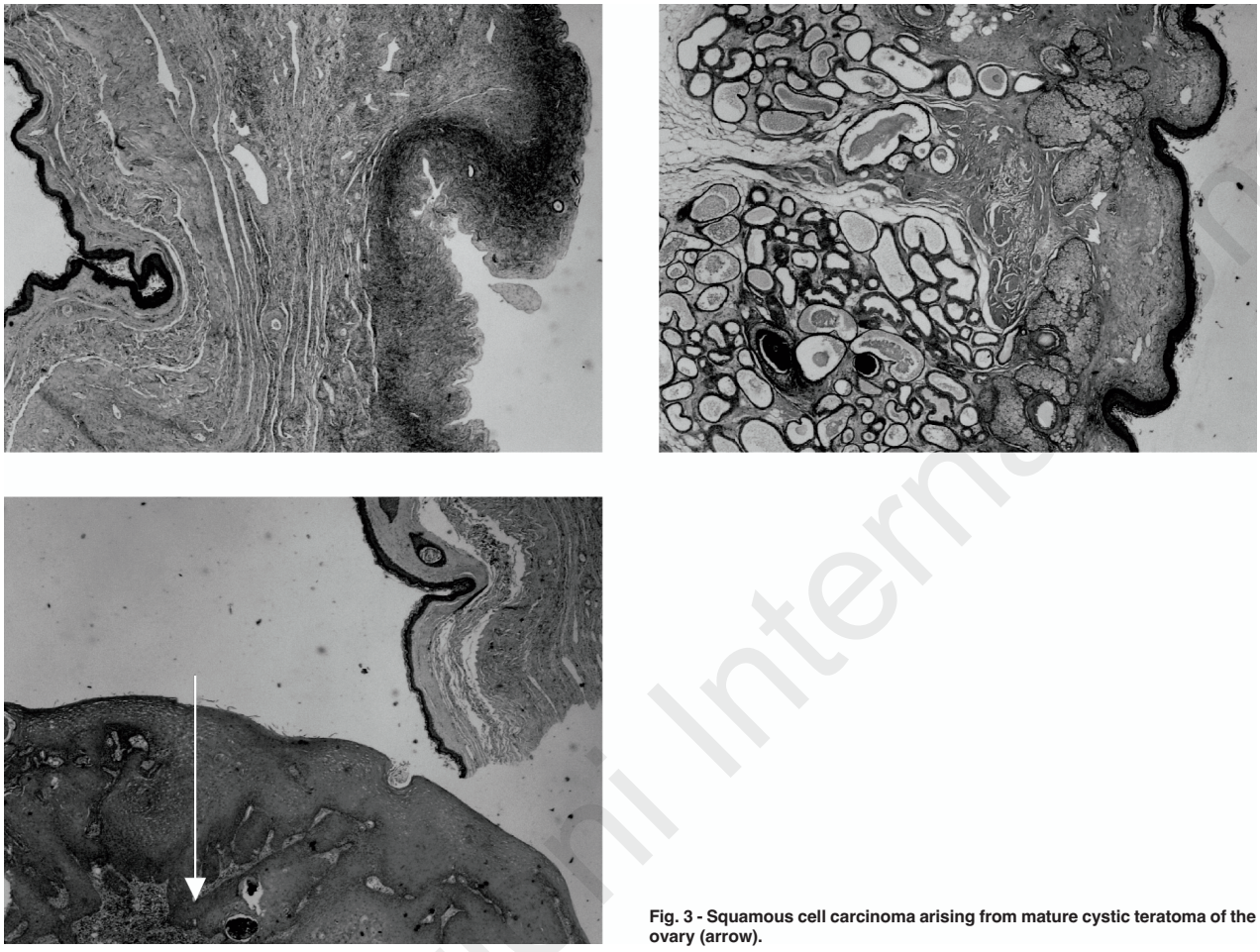

Fig. 3 - Squamous cell carcinoma arising from mature cystic teratoma of the ovary (arrow).

ratinising squamous epithelium with marked acanthosis and atypia, reminiscent of differentiated VIN-III and superficially SCC. The lesion measured 1.2 cm. The external surface of the cyst was intact (Fig. 3).

Discussion

Malignant transformation of MCTs may arise from any of three germ cell layers present in the teratoma, with an average frequency of 1-2% (12). This rare malignancy may occur at any age and it is detected most often in postmenopausal women with a median age of occurrence between 45-60 years old (1, 4). It is of great importance to know that MCTs arising in patients older than 45 years old sustain a higher suspicion of malignancy (13). In our case, woman's age was 56 years old and it is worth to mention her premenopausal status.

On the grounds that MCT is a common neoplasm diagnosed most usually accidentally in a routine pelvic examination checkup or during pelvic ultrasound scanning after atypical patient's symptoms, it has been recently given great importance in the preoperative risk evaluation of this neoplasms in order to improve surgical and therapeutic outcome. Patient's age older than 45 years, tumor size and tumor growth rate, imaging characteristics and serum tumor markers are risk factors for malignancy in MCTs (12, 14). Gastrointestinal symptoms, rectal bleeding, or urinary frequency are various symptoms described and related to MCTs and they are attributed to invasion of nearby organs (4). The mean size of MCTs is 6-9 cm compared to malignant transformed MCTs which tend to have a larger size with an average mean size of 15cm (15, 16). Kikkawa et al. reported that a tumor size greater than 9.9 cm has a 86% sensitivity for malignancy (13). In our case the patient presented with abdominal pain and further evaluation with transabdominal and transvaginal ultrasound revealed a 10 x 6.27 cm pelvic mass containing fat, soft tissue and calcification.

SCC is the most common type of this rare pathologic condition followed by melanoma , adenocarcinoma,

sarcoma, carcinoid and thyroid carcinoma (17). In most cases it is not diagnosed preoperatively (16, 18, 19). The use of tumor markers as a diagnostic tool to discriminate MCTs and SCCs arising from an MCT is difficult to help since most of them are raised in both situations (2). However, CEA, squamous cell antigen, CA-125 and CA 19-9 levels have found to be statistically significantly higher in cases of SCCs rising from MCTs than MCTs (20-22)[Parithivel, 2011 #11;Miyazaki, 1991 #63;Tseng, 1996 #75]. In our case, tumor marker levels were normal.

The use of magnetic resonance and CT imaging in preoperative diagnosis is doubtful whereas most studies concluded that Doppler detection method is a more powerful indicator than serum squamous cell antigen levels (23, 24). A cystic mass containing fat or a heterogenous soft tissue mass (that possibly contains calcification elements) are classic diagnostic findings for MCTs (25). Malignant imaging characteristics include thick walls, enhancing solid components or papillary projections within the cyst, peritoneal deposits or lymphadenopathy (21). As we have already described, in our case transabdominal ultrasound showed a 6,27 x 10 cm pelvic mass containing fat, soft tissue and calcification elements whereas a subsequent CT of the abdomen and pelvis demonstrated a right pelvic mass measuring 4x6x10 cm without the presence of pelvic nodes, ascites or other malignant imaging features.

As a result of the rarity of SCC arising in MCT, there is no standard treatment and most often patients are treated in the same way with those patients with epithelial ovarian cancer (2). Multiple case series recommend surgery including total hysterectomy, bilateral salpingo-oophorectomy, omentectomy and pelvic-paraortic lymph node dissection with further platinum-based agent chemotherapy (2, 26). The role of radiotherapy still remains unclear (2, 26). In our case, after the final pathologic specimen result, we proceeded to a second surgery including total hysterectomy, left salpingo-oophorectomy, omentectomy and pelvic node dissection.

Conclusion

A rare and unusual disorder is SCC arising in an MCT. As an exceedingly rare disease there is no standard of therapy, with most cases progressing to surgery followed by chemotherapy.

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