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Carcinoid tumor arising from ovarian mature cystic teratoma with multiple intra-abdominal parasitic cystic teratomas: a new case report and literature review

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Summary. Mature cystic teratoma (MCT) is the most common type of ovarian germ cell tumor and is typically benign, but rare malignant transformation has been reported at a rate of 1~2%. Among a wide variety of malignant tumors deriving from an MCT, malignant transformation into carcinoid tumor is extremely rare and is not easy to diagnose preoperatively. Parasitic dermoid cysts, named after the autoamputation and subsequent reimplantation of an ovarian dermoid cyst elsewhere in the abdomen, are also exceedingly rare and their coexistence with a carcinoid tumor has not yet been reported. In this study, we report a new case of ovarian carcinoid tumor arising from a MCT with multiple intra-abdominal parasitic cystic teratomas.

Key words: mature cystic teratoma, parasitic cystic teraoma

Introduction

Mature cystic teratoma (MCT) is the most common type of ovarian germ cell tumor, accounting for 10-20% of all ovarian tumors during the female reproductive period. These tumors are typically benign, and rarely undergo malignant transformation, the incidence rate being 1~2% (1, 2).

The most common malignant tumor arising from a MCT is squamous cell carcinoma, whereas carcinoid tumors are extremely rare and not easy to diagnose preoperatively (1, 3, 4).

Parasitic dermoid cysts are also very rare and their coexistence with carcinoid tumors has not yet been reported (5, 6).

In this study, we report a new case of ovarian carcinoid tumor arising from a MCT with multiple intra-abdominal parasitic cystic teratomas.

Case report

A 50 year-old premenopausal woman, gravida 1 para 1, was referred to our department owing to a pelvic mass that was detected incidentally during a routine examination.

The patient had no significant past medical history but did have a surgical history of prior myomectomy conducted 25 years ago in Vietnam.

On pelvic examination, a large, non-tender cystic mass could be palpated beyond the umbilicus. Ultrasound examination (Fig. 1A) revealed multiple uterine myomas (the largest myoma being 5.5 cm in diameter) with a large multiseptated abdominopelvic mass (20 × 9 cm) containing highly echogenic mural nodules, multiple calcifications, and a complex hyperechoic right adnexal mass (7.6 × 7.2 cm).

Further testing of the patient was ordered, includ-

ing serum tumor marker levels (CEA, CA 19-9, CA 125, AFP) and magnetic resonance imaging (MRI) of the abdomen and pelvis. All serum tumor markers were within the normal limits, with the exception of elevated CA 125 (494.2 U/mL).

MRI of the abdomen and pelvis (Fig. 1B) showed a large multiple loculated mass (6.7 × 10.8 × 20.0 cm) with an internal enhancing mural nodule probably deriving from the left adnexa, a right adnexal mass (7.0 × 6.2 cm) with fat-fluid levels, and multiple omental masses (the largest mass being approximately 8 cm in diameter) which were suspected of being metastatic lesions.

Open surgical exploration was performed and a large, complex, multi-cystic tumor was found that appeared to have originated from the left ovary, as well as a solid-cystic right ovarian tumor, multiple parasitic masses attached to the great omentum, and multiple small nodules in the peritoneum.

Total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed and the masses in the omentum and peritoneum were carefully dissected and removed.

Frozen sections from the bilateral ovarian tumors and omental masses showed a mature cystic teratoma and no other intervention was performed at the time of surgery.

Macroscopic examination revealed a left ovarian cystic mass (19.0 × 11.0 × 10.0 cm) weighing 141.4 g that contained sebum and hair with solid components, a right ovarian cyst mass (5.5 × 2.4 cm) filled with yellowish sebum like materials, and omental cystic masses (9.3 × 7.0 × 6.5 cm) containing brownish serous and mucoid materials.

Microscopically, the right ovarian tumor was composed of thyroid follicle (Fig. 2A, 2B), squamous epithelium and sebaceous gland structures (Fig. 2C, 2D). Carcinoid tumor components were found in the left ovary with anastomosing ribbon-like nests (Fig 2E) and this was confirmed by a positive immunohistochemical (IHC) signal for synaptophysin (Fig. 2F) and chromogranin (Fig. 2G) and negative for CD 56 (Fig. 2H) .

The final pathological diagnosis was malignant transformation into a carcinoid tumor from MCT in the left ovary and multiple benign mature cystic teratomas from the contralateral right ovary, omentum and peritoneum.

The patient was discharged on the 8th postoperative day following an uneventful course.

We planned concurrent chemoradiation therapy following surgery; however, the patient was lost to follow-up.

Discussion

Mature cystic teratoma with secondary malignant transformation

MCT is the most common type of ovarian germ cell tumor and is composed of well differentiated tissues from all three germ cell layers (ectoderm, mesoderm, endoderm) (1, 2).

Malignant transformation of MCT is rare and occurs in 1~2% of cases, and is usually diagnosed postoperatively, by histology studies (3, 7).

Although malignant transformation can occur from any of the embryonic germ layers, the most common are squamous cell carcinomas while other carcinomas reported in association with MCT include: adenocarcinoma, basal cell carcinoma, adenosquamous carcinoma, thyroid carcinoma, sebaceous carcinoma, malignant melanoma, sarcoma, carcinoid tumor, and neuroectodermal tumor (2-4, 7).

Malignant transformation into carcinoid tumor is extremely rare, comprising an estimated 0.3% of all ovarian neoplasms, and is usually seen in postmenopausal women, apart from the mucinous carcinoid tumors typically seen in younger women (8).

Rarely, ovarian carcinoid tumors may secrete neurohormonal peptides that have been associated with carcinoid syndrome, a syndrome of hormonal excess, with symptoms including flushing, diarrhea, and bronchospasm (1).

In this case, a carcinoid tumor was found in the left ovary and the patient did not present with any features of carcinoid syndrome.

The prognosis in primary ovarian carcinoid is very good when treated with a radical surgical approach with en-bloc resection, whereas metastatic carcinoids have poor outcomes (8, 9).

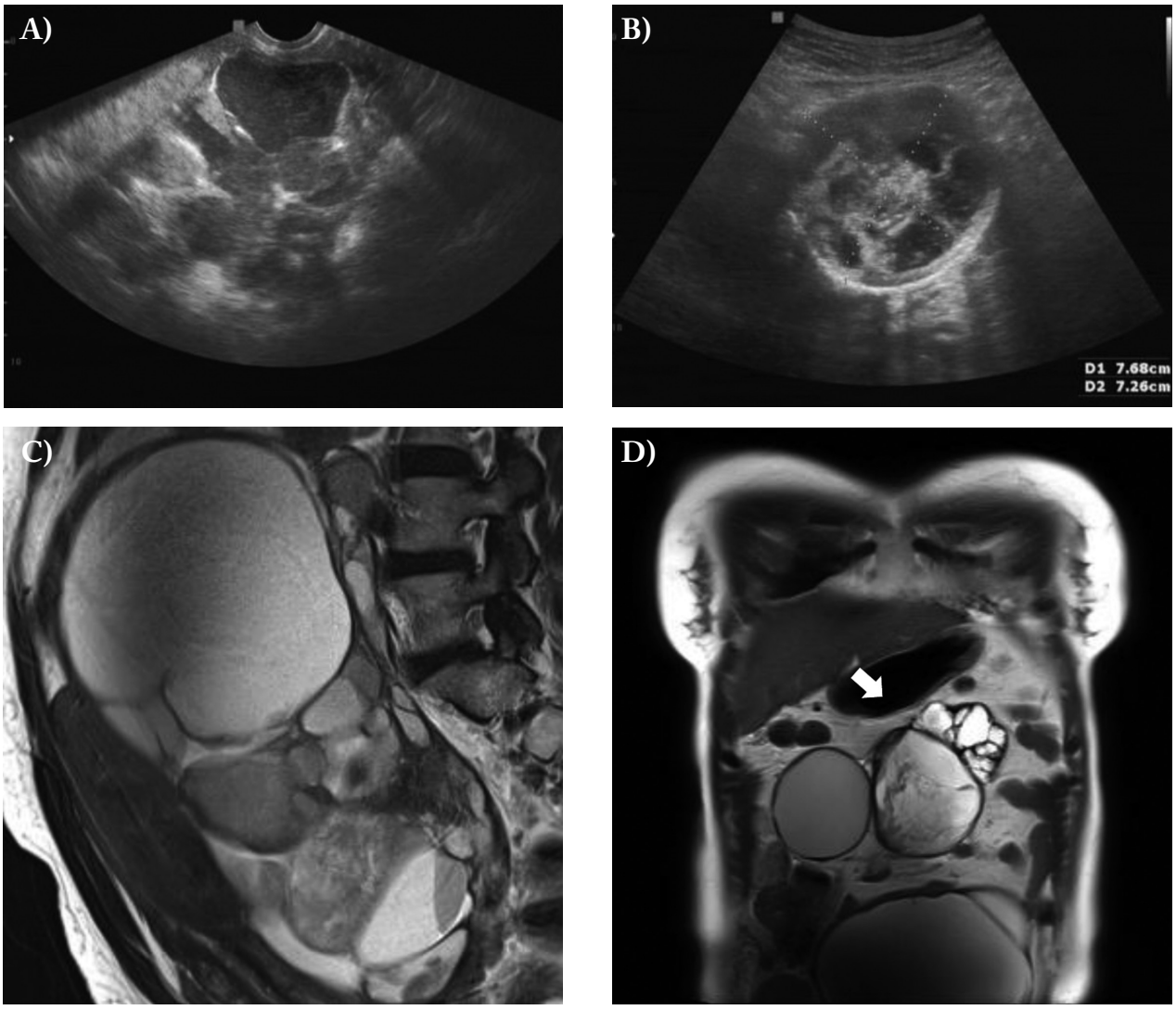

Figure 1. Transvaginal ultrasonography. (A) A large multiseptated abdominopelvic mass (20 X 9 cm) with multiple calcifications. (B) a complex hyperechoic right adenxal mass (7.6 X 7.2 cm).
Magnetic resonance imaging (MRI) of the abdomen-pelvis: (C) a large multiple loculated heterogenous mass with an internal enhancing mural nodule (6.7 x 10.8 x 20.0 cm). (D) multiple masses in omentum (white arrow) and pelvic cavity on T2-weighted MR images.

Parasitic omentalteratoma

Parasitic omental teratoma is an exceedingly rare tumor that usually originates from an ovarian dermoid cyst for which torsion, autoamputation and subsequent reimplantation into the omentum are performed (5, 10).

A review of the literature on parasitic demoid cysts indicates that the great omentum is probably the main location for secondary implantation of the tumor (10, 11).

The pathogenesis of omentral teratomas is unclear, but Ushakov et al. (5) reviewed three proposed etiologies of omental teratomas.

The first etiology is a primary omental teratoma. According to this theory, migration of germ cells along the primitive gut to the genital ridge may be arrested at the level of dorsal mesentery, which ultimately be-

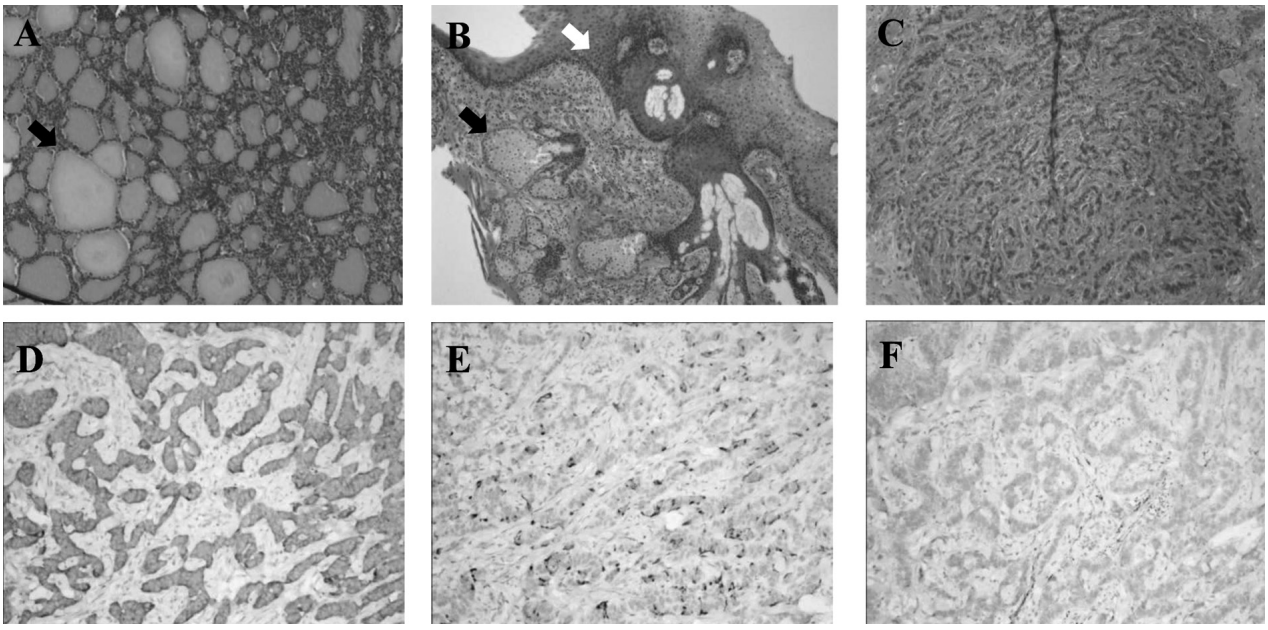

Figure 2. Microscopic view. (A) Hematoxylin and eosin staining: thyroid follicle structure (black arrow) in right ovary (X100) (B) Hematoxylin and eosin staining: skin structure with squamous epithelium (white arrow) and sebaceous gland (black arrow) in right ovary (X40) (C) Hematoxylin and eosin staining: carcinoid tumor with ribbon-like arrangements in left ovary (X200) (D) Synaptophysin positive in left ovary (X200) (E) Chromogranin positive (focal) in left ovary (X200) (F) CD 56 negative in left ovary (X200).

comes the greater omentum. However, no histological evidence for omental ectopic germ cells has ever been found.

The second etiology, suggests that the tumor may arise from a supernumerary ovary located in the omentum, though this is a rare gynecologic anomaly.

The third etiology, which is generally accepted for most cases, is autoamputation of an ovarian dermoid cyst and subsequent reimplantation in the greater omentum, because of the association between mature teratoma of the greater omentum and pathological evidence of ovarian stroma.

In this case, a large ovarian tumor may have undergone autoamputation and then reimplantation in the omentum and peritoneum.

Management of mature omental teratomas consists of local excision, with partial omentectomy (5).

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