Primary carcinoid tumor of the ovary arising in a mature cystic teratoma: a case report

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Summary

Primary ovarian carcinoid tumors are rare entities, they may appear with other teratomatous components, and can be often being mistaken as part of mature cystic teratomas. Consistent with their rarity and low incidence, imaging clues that could have led to suspicion of this tumor are not well-documented. Herein, the authors present a rare case of primary ovarian carcinoid tumor in a mature cystic teratoma, who initially presented with complaints of abdominal distension for months. Contrast-enhanced computerized tomography (CT) demonstrated a multilobular mass with different density components including fat, soft tissue, and calcification materials, as well as rich vascular supply from the right ovarian vein. Serum tumor markers were within normal limits. Bilateral salpingo-oophorectomy was performed and the pathological diagnosis was mature cystic teratoma with coexisting primary ovarian carcinoid tumor, insular type. The patient has remained well with no residual disease for over one year of follow-up.

Key words: Carcinoid tumor; Computerized tomography; Teratoma.

Introduction

Ovarian carcinoid tumors are rare, accounting for only 0.3 to one percent of all carcinoid tumors, and 0.1% of all ovarian neoplasms [1]. They may appear with other teratomatous components and can be often be neglected as part of mature teratomas [2]. Few women may present with teratomatous ovarian masses that are not properly diagnosed as carcinoid tumor until the time of histopathological analysis. Prior knowledge of correlated images of ovarian carcinoid tumor may be helpful to arouse alert of coexistent malignant component before operation. Nonetheless, literature that described detailed image findings of primary ovarian carcinoid tumors is sparse [3-8]. Herein, the authors present a rare case of primary ovarian carcinoid tumor in a mature cystic teratoma, and describe its detailed findings on computed tomography (CT).

Case Report

A 72-year-old woman experienced abdominal distension with bearing down sensation for several months. Ultrasonography showed a huge heteroechoic cystic pelvic tumor with acoustic shadows. Contrast-enhanced CT showed a right ovarian multilobulated mass with rich vascular supply from an engorged and tortuous right ovarian vein (Figure 1). Solid parts with miniscule streaks of vascular enhancement, calcification, and fat density were also found within the tumor (Figure 2). Serum tumor markers were within normal limits (carcinoembryonic antigen, 2.7 ng/ml; cancer antigen 125, 19.66 U/ml).

Exploratory laparotomy revealed a right well-circumscribed ovarian tumor (11 cm × 9 cm × 9 cm) with solid and cystic parts. The cystic part contained greasy, sebaceous-like material, and hair- and teeth-like structures, whereas the solid part showed tan-brown speckles with focal hemorrhage but no visible necrotic change (Figure 3). Owing to its mature teratomatous appearance, bilateral salpingo-oophorectomy was performed. The uterus and the left ovary were atrophic. Histopathological examination revealed right ovarian mature cystic teratoma with coexisting primary ovarian carcinoid tumor of the insular type (Figure 4). The carcinoid tumor cells stained positive for cytokeratin, synaptophysin, and chromogranin A. The patient has remained in good health without tumor recurrence at one-year follow-up.

Discussion

The appearance of ovarian carcinoid tumors on various imaging modalities is not characteristic enough for easy distinction from other solid tumors. Takeuchi *et al.* described ovarian carcinoid tumor as a solid component of homogenous soft tissue enhancement on CT [3]. On contrast-enhanced CT scans, the hypervascularity of carcinoid tumors can be observed as intense enhancement [3]. The isoattenuating density of the ovarian solid part compared to that of the psoas muscle, right tortuous and engorged ovarian vein (Figure 1), and miniscule streaks within the solid part (Figure 2) found in the presented case may represent the clinical characteristics of hypervascularity, indicating malignant ovarian tumor.

In the authors' knowledge, only a few case reports described their vague imaging findings of coexistent ovarian carcinoid tumors in mature cystic teratomas (Table 1) [5-8]. Most authors described their ovarian neoplasms with calcification and fat contents. Only one case report mentioned about adnexal mass with the solid component, which was later confirmed as the carcinoid tumor [5].

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Table 1. — *Clinical features of patients with an ovarian carcinoid tumor in a mature cystic teratoma.*

Case No.	Authors	Age (years)	Clinical presentations	Image findings	Management	Follow-up period	Outcome
1	Guney *et al.* [5]	54	Abdominal fullness, abnormal uterine bleeding	US: right ovarian cystic mass with solid components CT: right adnexal mass of solid and cystic components with calcifications	TAH, BSO	3 months	NED
2	Torre *et al.* [6]	76	Abdominal fullness	US: left ovarian tumor CT: left ovarian tumor with invasion to the uterine serosa	TAH, BSO, Pelvic LN dissection	70 months	NED
3	Kester *et al.* [7]	25	Urinary frequency, urgency, nocturia	CT: a left ovarian mass with fat and calcification	Left oophorectomy	NA	NED
4	Ansell *et al.* [8]	54	Bilateral ankle edema, elevated jugular vein pressure, systolic murmur	CT: dermoid cyst-like ovarian mass	TAH, BSO, mesenteric LN biopsy, appendectomy	8 months	NED

†BSO: bilateral salpingo-oophorectomy; CT: computerized-tomography; LN: lymph node; NA: not available; NED: no evidence of disease; TAH: total abdominal hysterectomy; US: ultrasound.

Fig. 1

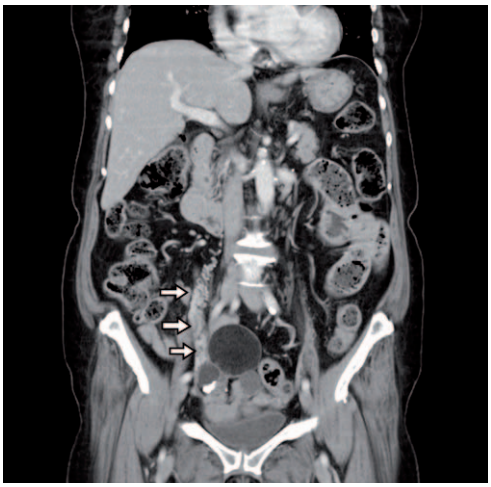

Fig. 2

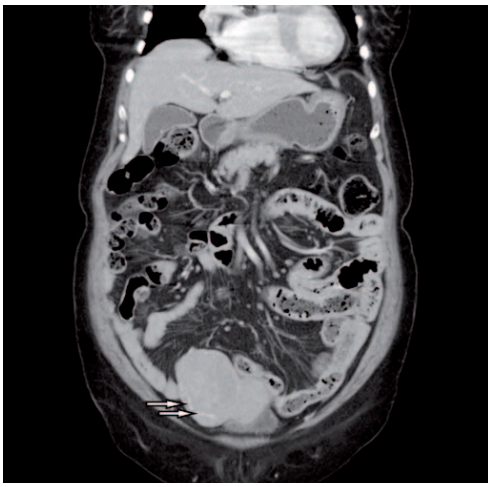

Fig. 3

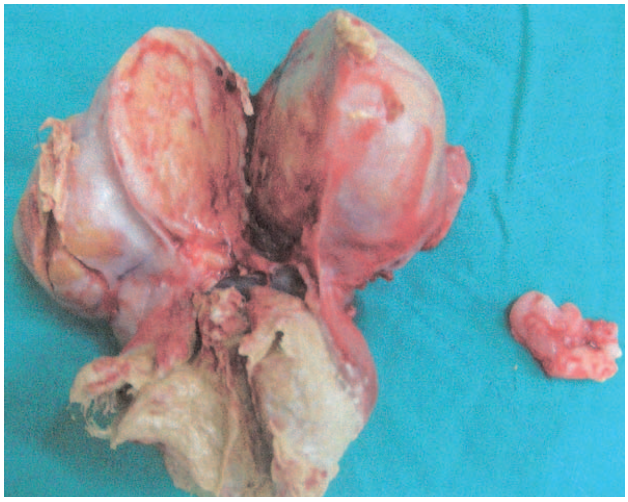

Fig. 4

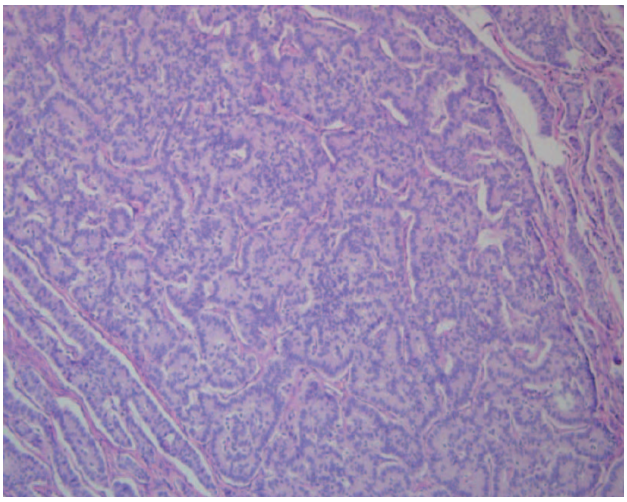

Figure 1. — Computerized tomography revealed a multilobulated mass with different density components. The tortuous and engorged ovarian vein (white arrows) courses along the right ureter and eventually ends in the tumor mass.
Figure 2. — Miniscule streaks of vascular enhancement (white arrows) are seen within the solid component.
Figure 3. — Right ovarian tumor with a whitish-yellow solid component and a cystic, greasy component.
Figure 4. — Histopathological examination of the right ovarian carcinoid tumor revealed uniform, monotonous tumor cells arranged in an organoid manner, insular to focally trabecular patterns (H&E, x100).

Only one-third of the insular type ovarian carcinoid tumors are symptomatic with flushing, diarrhea, bronchospasm, or dyspnea [9]. Therefore, diagnosis of carcinoid tumors based on symptom presentations is not reliable.

Although an ovarian tumor with hair, bone, and sebaceous contents is usually believed to be a mature teratoma; however, any concomitant ovarian cystic tumor with solid component and/or hypervascularity on CT scan should prompt the consideration of intraoperative frozen section examination, which was reported to be useful in determining the nature of any suspicious ovarian lesions with a good sensitivity and specificity of 91.2% and 98.6%, respectively [10]. Despite the good prognosis of ovarian carcinoid tumor, intraoperative confirmation of carcinoid tumor can prompt immediate gynecologic staging surgery without delay [11]. Aggressive debulking surgery is advised, followed by standard post-operative work-up including gut hormone analysis and radionuclide scintigraphy with Octreoscan [12].

Conclusion

In conclusion, carcinoid tumor and ovarian mature cystic teratoma may exist concomitantly, and prompt intra-operative investigation (such as frozen section examination) should be performed in cases of ovarian tumor with solid component and hypervascularity on CT scan despite their mature teratomatous appearance.

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