Ovarian strumal carcinoïd with mucinous cystadenoma component: a case report with literature review

Fong Tsun, M.B.C.h.B., Tseung Kwan O Hospital

Introduction

Strumal carcinoïd is a type of germ cell tumour characterized by the presence of thyroid tissue closely admixed with carcinoïd. It is the second most common type of primary ovarian carcinoïds (~40%) (1). The presence of glands or cysts lined by columnar mucinous epithelium is common (46%) but this component is often small and microscopically detected. Cases that have prominent mucinous component in which the diagnosis of mucinous cystadenoma is warranted are rarely reported (2,3). Herein I report a case of ovarian strumal carcinoïd with mucinous cystadenoma component, followed by review of current literature on this entity.

Case presentation

Clinical findings

A 48-year-old woman with history of appendectomy was presented to the surgical unit of our hospital for left lower abdominal pain. Blood tests were unremarkable except mildly elevated white blood cell count (13.8x10^9/L). CEA and CA125 were within normal levels. Diverticulitis was the initial diagnosis. Her symptoms subsided after antibiotic and symptomatic treatment. She was discharged with CT scan of abdomen and pelvis referred. She had CT scan in private sector which revealed a mildly lobulated, hypodense and heterogeneous enhancing mass (3.8 x 5.1 x 4.6 cm) with focal cystic component in the right adnexal region (Fig. 1). Radiological differential diagnoses included right ovarian tumour, complex ovarian cyst or endometriotic cyst. She was referred to the gynaecological unit and bilateral salpingo-oophorectomy was performed.

Macroscopic findings

Intraoperatively, a 6 cm x 5 cm multiloculated mass with smooth intact capsule was noted in the right ovary. The specimen was ruptured in the specimen bag during retrieval with mainly solid material and small amount of mucinous content remained. The specimen we received consisted of a right fallopian tube (5 cm in length and 0.5 cm in diameter) attached to a light red to yellow mass (3 x 3 x 3 cm) and multiple light red to yellow tissue fragments (7 cm x 6 cm x 2 cm in aggregates). Cut surface of the tissue fragments showed focal gelatinous tissue. The left fallopian tube and ovary were grossly normal.

Microscopic findings

The tumour showed a thyroid component with colloid containing follicles and a carcinoïd component with mainly trabecular pattern. The carcinoïd cells showed stippled chromatin and eosinophilic granular cytoplasm. Mitosis is inconspicuous. A cystic mucinous component composed of cystic spaces lined by basolateral glandular epithelium with goblet cells was also noted, in keeping with mucinous cystadenoma. No significant atypia is seen in the thyroid, carcinoïd or mucinous component. No somatic malignancy or other germ cell tumour component is seen.

Immunohistochemical findings

The findings of immunohistochemistry are summarized as follows:

<table>
<thead>
<tr>
<th>Component</th>
<th>Strumal component</th>
<th>Carcinoid component</th>
<th>Mucinous component</th>
</tr>
</thead>
<tbody>
<tr>
<td>Synaptophysin+</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chromogranin+</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>SATB2+</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CDX2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Thyroglobulin+</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>TTF1+ (f)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CK7+ (f)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CEA</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CA125</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Diagnosis and follow-up

The case was signed out as ‘strumal carcinoïd with cystic mucinous component’ and follow-up was advised. The patient had an uneventful post-operative course and discharged on post-operative day 1. Follow-up at 3 months showed no evidence of recurrence and progression.

Discussion

Strumal carcinoïd usually occurs in postmenopausal women, but the incidence age ranges from 21 to 77 years (4). It can present as an incidental finding or with signs and symptoms related to the mass. Uncommonly it can secrete a wide range of neuro-hormonal peptides with different clinical presentations such as carcinoid syndrome (serotonin secretion), chronic constipation (peptide YY secretion) (5) and hypoglycemia (insulin secretion) (6).

Grossly, strumal carcinoïd is usually unilateral and appears as a solid mass with variable cystic content. Cut surface is usually yellow or tan, and may show gelatinous tissue if mucinous component is present, as in this case. Dermoid cyst component can also be present.

Microscopically, the tumour is composed of two components – struma ovarii and carcinoïd and they are intimately admixed. The carcinoïd component is usually trabecular (as in this case), but insular or mixed pattern can occur (2). Mitotic activity is usually low, but a case with increased mitosis and necrosis has been reported (7). Teratomatous component may be present. Glands or cysts lined by columnar mucinous epithelium are present in 46% of the cases, and possibly represent common teratomatous origin from a pluripotential endodermal cell (8). In about 10% of the cases, the mucinous component is so prominent that a diagnosis of mucinous cystadenoma can be made (2,8,9) as seen in this case. Rarely mucinous cystadenocarcinoma has been reported in the mucinous component (10) and thyroid carcinoma (papillary/follicular type), has been reported in the strumal component (11). There are limited studies on the immunoprofile of strumal carcinoïd. The largest one comes from a case series of 13 cases (11). The immunoprofile in this case is largely similar to that case series, except that synaptophysin is diffusely positive in the thyroid component and thyroglobulin is negative in the carcinoïd component. Interestingly, SATB2, a marker for colorectal origin presumably, is diffusely positive in the carcinoïd and thyroid component in addition to the mucinous component.

Strumal carcinoïd usually has an excellent prognosis. Treatment ranges from unilateral salpingo-oophorectomy to bilateral salpingo-oophorectomy with or without total hysterectomy and is usually curative. Rare cases of lymph node metastasis, distant metastasis and death have been reported (12). The effect of the presence of mucinous component on prognosis is uncertain. It is also of note that a case of pseudomyxoma peritonei has been reported in a strumal carcinoïd with mucinous component (13). The mucinous component in that case was morphologically very similar to this case, signifying the potential aggressive behavior of this seemingly benign mucinous component.

Conclusion

Strumal carcinoïd can be rarely associated with a mucinous cystadenoma component. Aggressive behavior has been rarely seen in all of these three components. Follow-up is necessary in this situation.

References