Case Report

Primary juxtaovarian yolk sac tumor concurrent with an ipsilateral ovarian mature teratoma in an adult woman: a rare association

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Abstract: Objectives: Yolk sac tumor (YST) is a rare neoplasm that primarily occurs in the ovary in children and young women. Previously, it has been reported that the teratomatous components can be recognized in part of YSTs or appear in the contralateral ovary. Here, we report a rare case of an adult woman with a juxtaovarian YST concurrent with an ipsilateral ovarian mature teratoma. Methods: A 47-year-old woman found a pelvic mass for nine days and subsequently underwent debulking operation. The specimens were evaluated for detailed characterizations through gross examination, microscopy and immunohistochemistry. A literature review was performed and the pathogenesis was briefly discussed about the generation of an extraternal YST concurrent with a teratoma. Results: The right juxtaovarian tumor showed typical histological patterns of YST. Immunostaining demonstrated the YST nature of Gly-3 and AFP positive tumor cells. The ipsilateral ovarian tumor was a common mature cystic teratoma with chronic fibrotic changes. According to the differences of the origin and the differentiation of the two germ cell tumors, we suspect that the occurrence of the teratoma is earlier than the YST. Conclusions: To our knowledge, this is the first report of an adult woman with a juxtaovarian YST concurrent with an ipsilateral ovarian mature teratoma.

Keywords: Yolk sac tumor, extraovarian, ovarian mature teratoma, ipsilateral, concurrent, adult

Introduction

Yolk sac tumor (YST) is a rare malignancy that primarily occurs in the ovary in children and young women. The teratomatous components can be recognized in some YSTs. Benign cystic teratoma can appear in the contralateral ovary in about 10% of the cases. However, it is extremely rare to encounter a juxtaovarian YST concurrent with an ipsilateral ovarian mature teratoma in an adult woman. We present one such rare and interesting association of these two diseases.

Case report

A 47-year-old woman presented with menstrual disorders for six months and a pelvic mass for nine days. The laboratory tests showed an extremely high level of alpha-fetoprotein (AFP) at 1874.1 ng/ml (reference: < 13.4 ng/ml). Ultrasonography and Magnetic resonance imaging (MRI) findings were suggestive of ovarian malignancy in the right adenexa (Figure 1A, 1B).

Explorative laparotomy was performed. The mass was found near the right ovary and was located between the mesovarium and the middle of ipsilateral fallopian tube. The right ovary appeared slightly enlarge. The frozen section of the mass was suggestive of YST. Thus, a total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy and pelvic lymph nodes dissection were performed.

Pathological findings

The uterus, bilateral adnexas, greater omentum and pelvic lymphadenectomy specimens were sent separately.

On gross examination, an encapsulated mass measuring about 7 cm × 6 cm × 5 cm was discovered in the right juxtaovarian area focally attaching to the ipsilateral fallopian tube. The sectioned surface was solid, soft, gray and...
partly presented necrosis and hemorrhage. The right ovary measured about 2.5 cm × 2 cm × 0.8 cm, in which a cystic tumor measuring 1.0 cm in maximum dimension was present (Figure 2A, 2B). The left adnexa and uterus were unremarkable.

Microscopically, the right juxtaovarian tumor showed numerous histological patterns: a typical papillary, reticular (microcystic), glandular pattern and partly micropapillary structures with perivascular Schiller-Duval bodies (Figure 3A, 3B). In addition, hyaline globules were observed in some areas. The cystic tumor in the right ovary was composed of dermoid cyst components of a few hairs and small foci of well-differentiated hyaline cartilages (Figure 3C). In addition, multinucleated giant cells, calcification and significant fibrosis were seen in the capsule wall, which suggested that the tumor growth had gone through a long time. The malignant tumor was confined to the right juxtaovarian area. No metastases were found.

Immunohistochemistry of the juxtaovarian tumor revealed diffuse expression of AE1/AE3,
Gly-3, CD117, CD99, β-Catenin, P53 and scattered expression of AFP (Figure 3D, 3E), while CD30, HCG-β, CK7, CK20, WT-1, ER, PR, PAX-8, Inhibin-α, Napsin A were negative.

Histopathology confirmed a YST of the right juxtaovarian area concurrent with ipsilateral ovarian mature cystic teratoma.

Follow-up

On the thirteenth postoperative day, her AFP level decreased to 137.3 ng/mL, and on the twentieth postoperative day, it was 51.6 ng/mL.

Discussion

The yolk sac is an embryonal structure and also contains germ cells that migrate bilaterally to the urogenital crest during embryogenesis, where they will become the future gonads. Ovarian teratomas are the most common germ cell tumors generating in the yolk sac and most of them are mature and behave as benign tumors. The teratomatous components can also be recognized in part of YSTs. In 10% to 15% cases, germ cells with abnormal migration and trapped along the midline without degradation can result in extraovarian YST development, from the brain to the sacrococcygeal region [1-3]. It seems to be the most likely explanation for the extraovarian YST in our case.

More interestingly, benign cystic teratoma can appear in the contralateral ovary in about 10% of ovarian YSTs in the literature, while the teratoma is in the ipsilateral ovary in our case. According to the differences of the origin and
the differentiation of the two germ cell tumors, we suspected that the occurrence of the teratoma is earlier than the YST. Histopathology confirmed that the teratoma existed earlier in our case.

There were few reports about the recurrence of mature teratomas as YSTs. In a study by Yoshida et al, 13 cases of YSTs were found after sacrococcygeal teratoma (SCT) resection, which accounted for 5.4% of 241 SCTs patients enrolled [4]. Utsuki et al described a case of the malignant transformation of an intracranial mature teratoma into a YST in a 16-year-old boy [5]. D’Antonio et al reported a YST arising in the fallopian tube of an elderly woman. They speculated this YST could originate from a pre-existing (rare) mature teratoma [6].

It is worth noting that YST usually occurs in children and young women at the median age of 17.6 years, but the patient in our case is 47-year-old. As far as we are aware, this is the first case of a juxtaovarian YST concurrent with an ipsilateral ovarian mature teratoma in an adult woman.

YST has the ability to synthesize AFP. The significantly increased serum AFP level is of great significance in diagnosing YST and monitoring tumor recurrence in YST patients. Yoshida et al found that intervals of AFP measurement ≤ 4 months helped to detect subclinical localized YSTs for resection. They recommend that patients undergo serum AFP monitoring every 3 months for ≥ 3 years after SCT resection [4].

Surgical operation followed by cisplatin-based chemotherapy is always recommended for any YSTs [7].

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Disclosure of conflict of interest

None.

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