PRIMARY YOLK SAC TUMOR OF OMENTUM: A CASE REPORT

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Yolk sac tumor (YST) is one of the rare malignant germ cell tumor and usually occurs in gonad. Extragonadal sites of YSTs are reported in mediastinum, vagina, brain, and retroperitoneum but are extremely rarely in omentum. The clinicopathologic feature of primary omental YST is not well known and there are only 5 cases reported currently. Recently we experienced a primary YST of omentum in 27-year-old woman who was performed exploratory laparotomy due to abdominal distension and pain. She has remained free of diseases for 2 years with normal menstruation after the fertility-saving surgery and postoperative adjuvant chemotherapy with bleomycin, etoposide, cisplatin regimen. The subject of primary YST of omentum is reviewed, and the possible histogenesis of the tumor is discussed.

Keywords: Yolk sac tumor; Germ cell tumor; Omentum

Case Report

A 27-year-old woman, single of nullipara, was referred to department of gynecology due to aggravating abdomen distension and pain that develop incidentally a month ago. On history taking, her menstrual cycle was regular with 28 day cycle without any menorrhagia or dysmenorrhea. On physical examination, fetal head sized pelvic mass was palpated with a tenderness but without rebound tenderness. Laboratory test showed negative urine-human chorionic gonadotropin and no leukocytosis. Computerized tomography (CT) scan showed a large lobulating contoured pelvic mass infiltrating the omentum with ascites and a low attenuating nodule at right pericardiophrenic area (Fig. 1).

On the exploratory laparotomy, there was a 13 × 12 cm multiloculated solid, yellowish-grey mass in the greater omentum with multiple small nodules measuring 1-1.5 cm in greatest dimension. Another 7 × 5 cm white yellowish and friable multiloculated mass was found in cul-de-sac with small amount of ascites. The uterus, both fallopian tube and right ovary are grossly normal, but there was a 1.5 × 0.5 cm small nodule at left ovary. The frozen biopsy showed a malignancy favoring YST. In abdominal and pelvic cavity, disseminating bean sized small nodules were also found. Debulking surgery with the resection of left ovarian surface...
masses and the cul-de-sac mass, pelvic lymphadenectomy, omentectomy and resection and reanastomosis of the small bowel were performed saving the uterus and both ovary. Emergency laboratory test showed an elevated alpha fetoprotein (AFP) 6,065 U/mL. On pathological examination, the omental mass measured $13.5 \times 13 \times 7$ cm and 603 g weight. The cut surface is white, soft, hemorrhagic, and partly shows necrosis (Fig. 2).

Histological evaluation showed the YST exhibiting the Schiller-Duval body (Fig. 3A) and hyaline globules (Fig. 3B). Special immunohistochemical staining of the tumor showed positive for AFP (Fig. 3C), cytokeratin and negative for $\beta$-HCG.

On the 10th day after surgery, adjuvant bleomycin, etoposide, cisplatin (BEP) chemotherapy consisting of bleomycin (20 mg iv on days 2, 9, and 16), etoposide (100 mg/m² iv for 5 consecutive days), and cisplatin (15 mg/m² iv for 5 consecutive days) was begun. A total of four cycles of chemotherapy were repeated every 3 weeks. The patient showed complete response after 6 cycles of chemotherapy and showed no evidence of disease on physical examination, serum AFP level, and abdomino-pelvic CT scan for 2 years.

**Discussion**

The extragonadal YST is rare malignant germ cell tumor arising outside ovary or testes. The most common site of extragonadal YST are reported in sacrococcygeal region followed by anterior mediastinum, vagina, vulva, cervix, and uterine corpus [4]. However, primary YST of omentum is extremely rare and only 5

![](image1.png)

**Fig. 1.** Computerised tonography showed a large lobulating abdominal mass infiltrating the omentum with small amount of ascites.

![](image2.png)

**Fig. 2.** Gross feature of specimen showed a multinodular omental mass measuring $13.5 \times 13 \times 7$ cm. The cut surface is white, soft, hemorrhagic, and partly necrotic.

![](image3.png)

**Fig. 3.** Histological evaluation of the specimen shows typical Schiller Duval bodies (arrows) in microcystic area (A, H&E, $\times 200$) and hyaline globules (arrows) (B, H&E, $\times 400$). Special immunohistochemical staining of the tumor showed positive for AFP (C, immunohistochemical staining, $\times 400$).
cases are reported currently [1,2,4-6].
Omental YST shows histologic characteristics similar to that of
gonadal YST such as microcystic, endodermal sinus, solid, alveo-
larglandular, polivesicular vitelline, myxomatous, papillary, mac-
rocystic, hepatoid, and glandular or primitive endoderm patterns
[5]. Schiff positive intracytoplasmic and extracytoplasmic globules
are typical features of YST and these globules are usually positive
immunohistochemically for AFP, A1AT, transferrin, and basement
membrane components such as fibronectin, type IV collagen,
vimentin, and laminin [4,5]. In our case, typical histologic charac-
teristics of YST such as Schiller–Duval bodies and positive immu-
nohistochemical staining for AFP and cytokeratin.
The histogenesis of extragonadal YST remains controversial. One
hypothesis is that these tumors arise from aberrant differentia-
tion of somatic cells, and the other one is that the germ cells have
been arrested in their embryonic migration. The arrested embryon-
ic migration hypothesis explains that when the primitive gonadal
ridge expanded to the region of the external genitals, some of
germ cell arrested anywhere along the migration course and can
be the possible site of germ cell tumor in the future [4]. Metastasis
from an occult focus in the ovary may contribute to primary YST of
omentum, but in our case, thorough pathologic examination has
excluded this possibility.
In this case, omental mass was huge main mass and left ovarian
mass was located at ovarian surface without invasion, grossly. Al-
though in pathologic examination small nodule at left ovary shows
yolk sac tumor, ovarian capsule infiltration was not confirmed.
Mass of ovary was one of metastatic seeding nodules as like those
disseminated in abdominal and pelvic cavity.
Generally, gonadal YST is a disease with a peak incidence at 20
years [5], however, the ages of in w primary YST of omentum
omen previous reported were is 37, 44, 45, and 46 years implicat-
ing a rather older women’s disease [2,4,6]. However, primary YST
of omentum in our report showed a general young age of gonadal
YSTs.
Because omental YST also affects women of child bearing age, the
treatment of ometnal YST may not different from that of gonadal
YST [7]. Any gross metastases should be resected if possible, and
“fertility preserving” procedure can be considered even when the
tumor may remain after surgery [3,8]. Preservation of uterus and
ovaries are strongly recommended because the high response
rate of chemotherapy with BEP [3,9,10]. In previous report, the
average age of the 4 women with a primary YST of omentum was
43 years and there was no need to ‘fertility-saving surgery’. Actual-
tly total hysterectomy was performed in all the 4 women with
primary YST of omentum [2,4,6]. However, in this report, we per-
formed fertility saving surgery consisting of removal of the pelvic
tumor, pelvic lymphadenectomy, omentectomy and resection of
abdominal metastases with preservation of the uterus and both
ovaries. Although the child bearing outcome is not confirmed yet,
this is the first report that fertility saving surgery was performed in
primary YST of omentum.
Primary YST of omentum responds well to combination chemo-
therapy [2,4,6]. A regimen of BEP has been a major advance in
the therapy of advanced and localized germ cell tumors [3]. Our
patient also showed complete response with 6 cycles of BEP
combination chemotherapy.
Although long term outcome of omental YST is not well known,
all the 5 patients with primary YST of omentum reported showed
a favorable outcome [2,4,6]. In this report, the patient showed no
evidence of disease on physical examination and serum a-fetopro-
tein level and imaging studies for 2 years without any menstrual
abnormality.
In conclusion, we report a 27-year-old woman with a primary YST
of omentum successfully with fertility saving surgery and postop-
erative adjuvant chemotherapy with a brief review of literature.

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복막에 발생한 난황낭종양의 1예

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난황낭종양은 생식 세포에 속하는 종양으로서 비교적 흔한 암종은 아니지만 악성도가 높다. 종양의 대부분은 생식 기관(난소, 고환)에서 기원하며, 난황낭종양의 약 10-15% 정도가 생식기관 이외의 위치에서 발생한다. 복막에서 난황낭종양이 발생한 경우는 매우 드물며, 현재까지 5명의 환자가 보고된 바 있다. 27살의 비교적 젊은, 본 경우의 환자는 복부팽만을 주소로 내원하였고 우리는 시험적 개복술과 수술 후 보존적 항암치료를 시행하였다. 수술은 향후 임신 가능성을 고려하려 자궁과 난소를 보존하는 방향으로 이루어졌다. 항암치료 종료 2년 동안 추적 관찰하였으며, 현재까지 검사 소견상 재발이나 전이의 어떤 소견도 보이지 않고 있다.

중심단어: 난황낭종양, 생식세포종양, 복막