Primary yolk sac tumor of the omentum: a case report and literature review

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Summary

Background: Yolk sac tumor (YST) is the second most common malignant ovarian germ cell tumor, while a YST arising in the omentum is an exceedingly rare malignancy. *Case:* A 44-year-old woman was admitted with a history of abdominal distension of a month's duration. The alpha-fetoprotein (AFP) serum level was elevated to 27,612 ng/ml, and CT scanning reported an obviously thickened omentum. Explorative findings revealed a large omental mass with a small implantation on the surface of the left ovary. Histological evaluation of the specimen after surgery exhibited typical patterns of YST, and the specimen was stained for AFP, cytokeratin, and placental alkaline phosphatase. The patient was treated with total abdominal hysterectomy with bilateral salpingo-oophorectomy and infracolic omentectomy followed by four cycles of bleomycin, etoposide, and cisplatin combination chemotherapy. She has remained free of the disease for seven months after completion of therapy. The subject of YST arising in the omentum is reviewed. *Conclusion:* This is the fourth case of primary omental YST in females. The case of omental YST must be seriously considered once the tumor shows omentum thickening with elevated AFP serum levels.

Key words: Yolk sac tumor; Omentum.

Introduction

Yolk sac tumor (YST), also known as endodermal sinus tumor, was first recognized as a distinct entity by Teilum [1]. It is a malignant germ cell tumor that simulates the yolk sac and is characterized by an elevated production of alpha-fetoprotein (AFP). Although most YSTs usually occur in the testis and ovary, about 20% manifest outside the genital tract, including in the mediastinum, sacrococcygeal region, cervix, vulva, pelvis, and retroperitoneum [2]. Omental YST is an exceedingly rare malignancy. To our knowledge, this is the fourth report of YST originating from the omentum in a female.

Case Report

A 44-year-old woman, para 1-0-7-1, visited the First Affiliated Hospital of Zhejiang University exhibiting abdominal distension symptoms of one month's duration. Her menstrual cycle was 30 days with normal flow and the last abortion had been ten years before. She denied any family medical history of malignant disease. On pelvic examination, the mass or uterus could not be palpated due to ascites. The patient's abdomen bulged, making it appear similar to a full-term pregnancy, and it felt painful under light pressure. Ultrasonography (US) showed a large amount of ascites and multiple low echo homogeneous solid masses on the pelvic wall. The largest mass was 3.3 cm in diameter (Figure 1). The uterus and adnexa had no enclosed mass. Computerized tomography (CT) scan revealed an obviously thickened omentum and nodular mass on the peritoneum, greater omentum, mesentery, intestinal surface, and left adnexa (Figure 2). The largest single mass was located in the greater omentum with a 3.9 cm diameter and showed highly heterogeneous enhancement after administration of contrast material. Scans of the liver, gall bladder, spleen, pancreas, kidney, bladder, stomach, uterus, and ovaries were normal. The AFP serum level was elevated to 27,612 ng/ml (normal < 10 ng/ml). The CA-125 serum level was 334.9 U/ml (normal < 25 U/ml), whereas the ß-HCG and carcinoembryonic antigen (CEA) levels were within the normal range. Cytology of a specimen of peritoneal fluid withdrawn under US control was positive for highly undifferentiated adenocarcinoma cells.

During exploratory laparotomy, a 23 cm tawny solid multilobulated mass, weighing 3100 g, was found in the greater omentum. Implantation foci with a 0.1-2.0 cm diameter could be seen on the pelvic peritoneum, mesentery surface, paracolic sulci, liver surface, and rectovaginal pouch (Figure 3). The right ovary and bilateral fallopian tubes were normal in size. The surface of the left ovary reflected implanted tumors in three areas with each tumor measuring 0.3 cm in diameter. The result of frozen biopsy from an omental mass was adenocarcinoma. Then, infracolic omentectomy, total hysterectomy, bilateral salpingo-oophorectomy, and cytoreductive surgery were performed with no macroscopic residual disease.

Under microscopic observation, tumor cells grew in different forms as adenoid, mamillary, or solid flake. Schiller-Duval bodies (Figure 4) could be seen, and many mitoses were observed. However, interfibrillar substance was rare. The tumor implanted on the left ovarian surface. Immunohistochemical studies showed cellular positivity for AFP, cytokeratin (CK), placental alkaline phosphatase (PAP), P53, and cell proliferation-associated antigen (Ki67). Staining of the epithelial membrane antigen (EMA), CK7, CK20, CD34, HCG, CA-125, vimentin, calcium binding protein (CBP), estrogen receptor (ER), and progesterone receptor (PR) were negative.

AFP serum value on the first day after surgery was 8944.3 ng/ml. Combined chemotherapy (BEP regimen) consisting of cisplatin (20 mg/m^2 for 5 consecutive days), etoposide (100 mg/m^2 for 5 consecutive days), and bleomycin (18 mg IM on days 2, 9, and 16) was given one week later every three weeks. Chemotherapy was repeated for four cycles. Before the second course of treatment, the AFP serum value dropped to 8.0 ng/ml. The patient was followed-up for seven months without clinical and radiological evidence of recurrence.

Revised manuscript accepted for publication April 8, 2010

Fig. 1

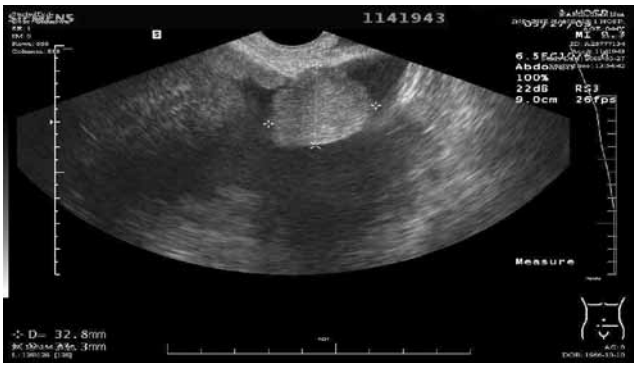

Figure 1. — Low echo occupations on the pelvic wall.

Fig. 2

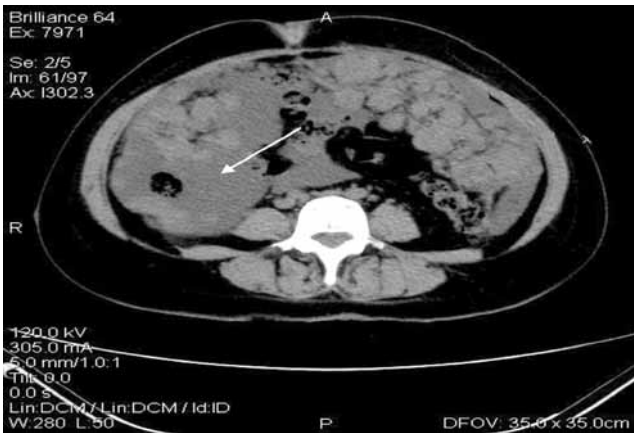

Figure 2. — Nodular foci in the greater omentum.

Fig. 3

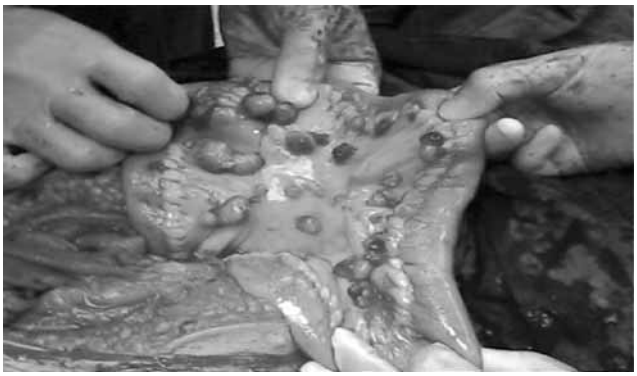

Figure 3. — Tawny tumor nubbles on the small intestinal mesenterium.

Fig. 4

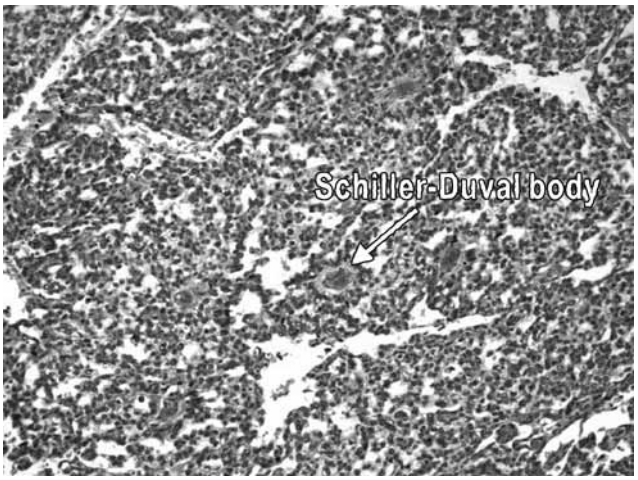

Figure 4. — Microscopic examination revealing reticular or papillary patterns of primitive germ cells with Schiller-Duval bodies (H&E × 200).

Discussion

YST is a highly malignant germ cell tumor that grows rapidly and metastasizes early through lymphatic and hematogenous routes [3]. It is the second most common malignant ovarian germ cell tumor after dysgerminoma and accounts for approximately 1% of all ovarian malignancies [4]. The case of omental YST is even more rarely reported. Reports that can be referred to presently consist of only three cases in females [5-7] and one of a 3-year-old boy [8]. The characteristics of these four cases in females are summarized in Table 1. Omental YST always occurs in females more than 35 years of age, with the average age being 43. The chief complaint is usually abdominal distension. The obviously thickened omentum and the ascites could be identified through CT or US.

The histogenesis of YST of the omentum remains speculative and controversial. One plausible hypothesis is that germ cells have been misplaced or arrested in their embryonic migration during embryogenesis and then become the potential tumor source [6]. In this case, the ovarian surface exhibited only small plantation foci. The greater omental pathological changes were obvious, and the pathological examination located no histological evidence of YST originating from the ovary. All this evidence supported the diagnosis of YST of the omentum.

The glycosidoprotein, AFP, is secreted by the embryo yolk sac or embryonal carcinoma cell with a serum half-life of five days. If the AFP value rises in adults, the diagnosis should first exclude hepatocellular carcinoma, cirrhosis, and hepatitis [9]. A positive rate of AFP serum elevation appeared in all omental YST cases. AFP serum level is a useful marker for the diagnosis and management of this kind of disease and is used to check for complete remission or recurrence. The prognostic value of a high AFP level at diagnosis remains controversial. Mayordomo *et al*. found that AFP > 1000 kU/l is associated with a higher risk of YST relapse [10], but Nawa *et al*. found that preoperative AFP serum levels before initial surgery have no significant correlation with prognosis [11]. In our case, the AFP value before surgery was 27612.3 ng/ml, which was the highest out of all the recorded AFP values of the omental YST cases. Although it dropped to a normal level rapidly after more than 40 days, long-term follow-up of the treatment effects require further observation. CA-125 was also found to have risen in our case, similar to the case studied by Kim *et al*. [5]. This increase may be due to peritonitis or infection accompanying the clinical features of abdominal pain.

Table 1. — *Clinicopathological features of primary omental YST in females*.

Author (Reference)	Age (yrs)	Chief complaint	Tumor marker (preoperation)	Frozen biopsy	Immunohistochemical stain positive	Immunohistochemical stain negative	Treatment	Chemotherapy (courses)	Follow-up
Kim *et al*. [4]	37	abdominal pain and distension	CA125: 374U/ml	Adenocarcinoma or mesothelioma	AFP, CK, PAP, vimentin, CK-7	HCG, CD 15, EMA	SCO, BSO, TAH, P&PA-LND, appendectomy	BEP (4)	1 year - NED
Park *et al*. [5]	45	abdominal distension	AFP: 20,250 ng/ml	YST	AFP, CK	vimentin, HCG,	ICO, BSO, TAH	BEP (4)	10 months-NED
Geminiani *et al*. [6]	46	abdominal distension	AFP: 21,550 ng/ml	Poorly differentiated neoplasm	AFP, CK, PAP, c-kit, vimentin, HCG, CD 30, CD 34, CD57, NSE, S-100 protein, PL EMA, SMA	chromogranin A, desmin	TO, TC, BSO, TAH-IS	BEP (6)	2 years - NED
Present case	44	abdominal distension	AFP: 27,612 ng/ml CA125: 334.9 U/ml	Adenocarcinoma	AFP, CK, PAP, P53, Ki67	vimentin, EMA, CK7, CK20, CD34, HCG, CA125, CBP, ER, PR	ICO, BSO, TAH	BEP (4)	7 months - NED

ICO infracolic omentectomy, *BSO* bilateral salpingo-oophorectomy, *TAH* total abdominal hysterectomy, *PD* poorly differentiated, *TO* total omentectomy, *TC* total colectomy, *IS* ileostomy, *NR* not reported, *WNL* within normal limit, *SCO* supracolic omentectomy, *P&PA-LND* pelvic and paraaortic lymph node dissection, *NED* no evidence of disease, *CK* cytokeratin, *PL* placental lactogen, *EMA* epithelial membrane antigen, SMA smooth-muscle actin, *CBP* calcium binding protein, *ER* estrogen receptor, *PR* progesterone receptor

The pathological diagnosis of YST is easily confused with that of clear cell carcinoma and adenocarcinoma [12]. In our case, the pathology before surgery was misdiagnosed as poorly differentiated adenocarcinoma. The typical YST has an embryonic structure similar to the fetal yolk sac, which performs a glomerular-like structure with a central vessel surrounded by prominent large cuboidal cells. Intracellular and extracellular hyaline globules were stained positive with periodic-acid-Schiff (PAS). Schiller-Duval bodies can be regarded as a diagnostic clue to YST in the presence of AFP, but they only exist in 20% of the YST cases. Immunohistochemical staining for AFP and CK was positive, while ER and PR staining were negative. These can be used for differentiating diagnoses. This article has presented a more detailed immunohistochemical study for YST of the omentum to help in the diagnosis of this disease.

Williams suggested that surgery combined with postoperative chemotherapy is presently the best mode of treatment for YST [13]. Three courses of BEP is the current standard therapy, and four courses are recommended in the case of bulky residual disease after surgery. The case of omental YST is very sensitive to this treatment. In all reported cases, the AFP level drops to the normal value before the second chemotherapy cycle.

This case reflects a rarely seen primary YST of the omentum and the patient is presently completely free of disease through effective surgery as well as four courses of BEP combined chemotherapy. Therefore, for tumors with no clear primary foci and with omental thickening, YST of the omentum must be seriously considered, and serum AFP testing can be helpful in differentiating the diagnoses.

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