Taiwanese Journal of Obstetrics & Gynecology 53 (2014) 409–412

Contents lists available at ScienceDirect

Taiwanese Journal of Obstetrics & Gynecology

journal homepage: www.tjog-online.com

Research Letter

Primary primitive neuroectodermal tumor of the ovary

Ling-Hui Chu [a], Wen-Chun Chang [a], Kuan-Ting Kuo [b], Bor-Ching Sheu [a, c, *]

[a] Department of Obstetrics and Gynecology, National Taiwan University Hospital and National Taiwan University College of Medicine, Taipei, Taiwan
[b] Department of Pathology, National Taiwan University Hospital and National Taiwan University College of Medicine, Taipei, Taiwan
[c] Centre for Optoelectronic Biomedicine, National Taiwan University Hospital and National Taiwan University College of Medicine, Taipei, Taiwan

ARTICLE INFO

Article history:
Accepted 27 August 2013

The most common neoplastic ovarian tumor in adolescence is the germ cell tumor. About 58% of primary ovarian neoplasms are germ cell tumors, and most are benign cystic teratomas [1]. Ovarian tumors composed of primitive neuroectodermal elements are extremely rare.

We report a case of an ovarian primitive neuroectodermal tumor (PNET) treated with fertility-sparing staging surgery and adjuvant chemotherapy, followed by six courses of chemotherapy.

A 16-year-old nulliparous girl presented with a pelvic mass which she had palpated accidentally. The mass grew rapidly over 3 weeks. The patient had experienced intermittent abdominal discomfort and irregular menstruation in recent months, but there had been no bowel habit changes. She visited our clinic and her tumor profile was checked. A detailed sonographic examination showed one huge, solid, 16.5 cm × 9.2 cm pelvic tumor with heterogenous echo complex contents including some cystic parts and calcification, with a suspected ovarian origin (Fig. 1). Abdominal and pelvic magnetic resonance imaging revealed a huge mass lesion in the pelvic cavity, which showed low T1 signal intensity and intermediate highT2 signal intensity, with some internal cystic components and good enhancement (Fig. 2).The patient was then referred to our oncology clinic, where tumor markers for suspected malignancy revealed an elevated carbohydrate antigen (CA)-125 level (120.9 U/mL), with normal levels of lactate dehydrogenase, carcinoembryonic antigen, CA-199, alpha-fetoprotein (AFP) and beta-human chorionic gonadotropin. The patient denied a family history of ovarian cancer and teratoma. She also denied symptoms of neuroendocrine activity such as flushing, diarrhea, abdominal cramping, and palpitations.

An exploratory laparotomy revealed a huge left ovarian solid tumor with an irregular border and central necrosis (Fig. 3). The uterus, bilateral fallopian tubes, omentum, and peritoneal surface were grossly normal. There was a moderate amount of ascites, about 400 mL. In consideration of her fertility at this young age, conservative staging surgery with a left salpingo-oophorectomy, dissection of left side pelvic lymph nodes, and infracolic omental excision was performed.

Immediate pathological frozen section revealed a spindle cell tumor. A detailed microscopic pathological examination showed the well-encapsulated ovarian tumor to be composed of grayish tan, solid, fleshy tissue with several small cysts with clear fluid. Marked necrosis was seen.

Microscopically, the tumor showed a high cellularity, composed of small cells with hyperchromatic, round to oval nuclei and scanty to small amounts of cytoplasm arranged in lobules separated by fibrovascular septa, patternless sheets incompletely divided by fibrovascular septa or a trabecular/cord-like pattern (Fig. 4). A fibrillary matrix was focally present. Mitotic activity (> 10/10 HPF, high power field) and apoptosis were frequently seen and tumor necrosis was evident. Immunohistochemically, the tumor was relatively diffusely positive for synaptophysin and cluster of differentiation (CD) 56, focally positive for chromogranin, S-100 and glial fibrillary acidic protein, with the latter two particularly predominant in the fibrillary matrix area, but negative for cytokeratin (AE1/AE3), CD99, and AFP. Based on the above findings, the tumor resembled a PNET of the central nervous system with a medulloblastoma/neuroblastoma pattern. In addition, small areas of mature teratoma composed of squamous epithelium, respiratory epithelium, bone, cartilage, smooth muscle, adipose tissue, mature glial tissue, and minor nondescript ducts were also present. Therefore, a primitive ovarian neuroectodermal tumor in association with a teratoma was considered.

The patient's postoperative course was smooth and she was discharged from the hospital after surgery. Under a diagnosis of left ovarian PNET, Stage IC, adjuvant chemotherapy for epithelial ovarian cancer was administered with the PT (carboplatin AUC: 6, paclitaxel 175 mg/m^2) protocol six times at 3-week intervals without severe side effects, except for alopecia.

She was regularly followed for 13 months at our clinic without evidence of recurrence, with regular menstrual cycles and a normal

* Corresponding author. Department of Obstetrics and Gynecology, National Taiwan University Hospital, Number 7, Chung-Shan South Road, Taipei 100, Taiwan.
E-mail address: bcsheu@ntu.edu.tw (B.-C. Sheu).

http://dx.doi.org/10.1016/j.tjog.2013.08.005

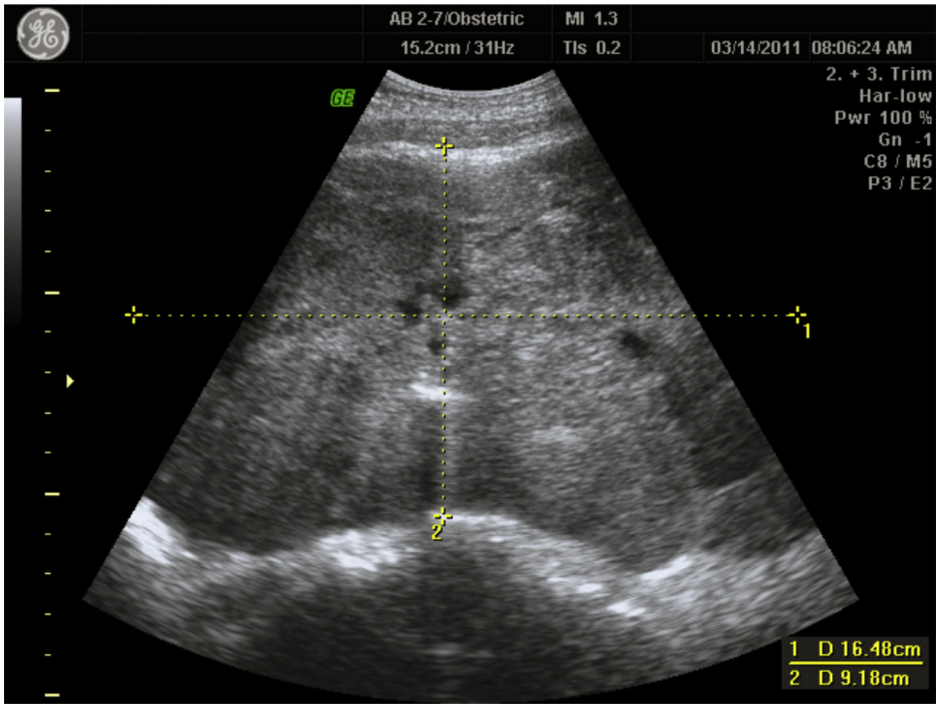

Fig. 1. Ultrasonography shows a huge, solid, heterogeneous tumor in the pelvic cavity.

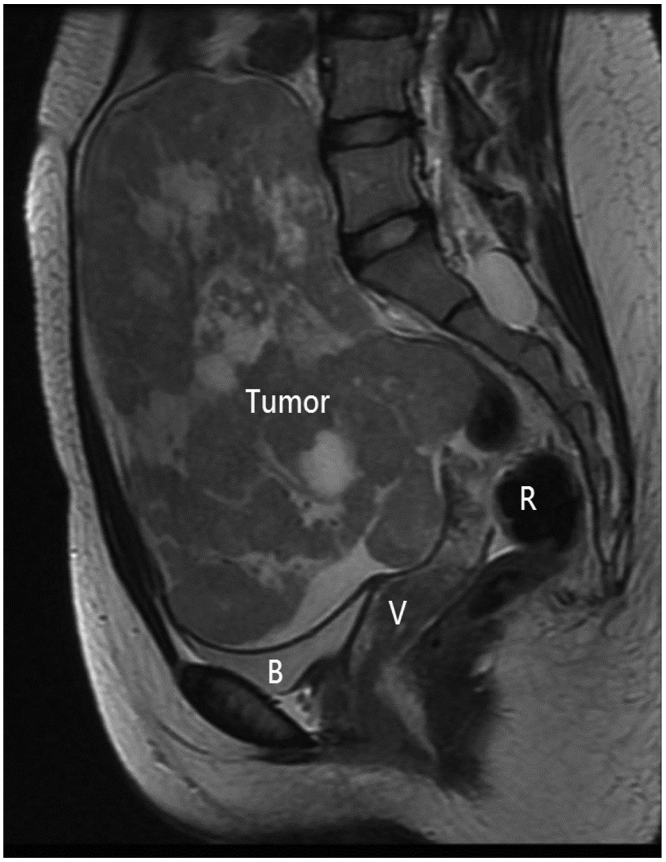

Fig. 2. Abdominal and pelvic magnetic resonance imaging reveals a huge mass lesion with low T1 signal intensity. B = urinary bladder; R = rectum; V = vagina.

hormone profile. The tumor makers including CA-125 were all normal during regular monthly follow ups. Follow up with fluorodeoxyglucose-positron emission tomography 12 months after completion of chemotherapy also showed negative findings.

Primary neuroectodermal tumors are rare monophasic teratomas composed of immature neuroectodermal tissue. The tumors are separate from other types of teratomas and the incidence is rare. These tumors are classified as ependymoma, astrocytoma, and primitive neuroepithelial tumors such as medulloblastoma, medulloepithelioma and neuroblastoma [2]. PNETs are highly cellular and composed of small cells with hyperchromatic, round to oval nuclei and scanty cytoplasm. Lobules separated by fibrovascular septa are present with prominent areas of necrosis [3].

Based on previous reports, most primary PNETs occur in the second to third decades of life, at a slightly younger age than the well-differentiated form of neuroectodermal tumors. The age range of patients in one study was 13 to 69 years [3]. The prognosis is generally poor with a high mortality rate.

Kleinman et al [3] reported 12 PNET cases in Stage IA to Stage III. The duration of posttreatment follow-up ranged from 2 months to 9 years. All patients with Stage IA-IC disease received unilateral salpingo-oophorectomy. Postoperative adjuvant chemotherapy was administered in three of four patients with Stage I disease. No patient had evidence of disease during follow-up. However, nearly all patients (7 of 8) in Stage III died of disease at 2–20 months after diagnosis.

Because of the rarity of cases, there is no consensus about the operative method or adjuvant therapies such as chemotherapy and radiotherapy. One case report in 2004 described a patient with PNET Stage IIIC with peritoneal carcinomatosis and extensive lymphadenopathy who received fertility-sparing staging surgery and adjuvant radiotherapy and chemotherapy with carboplatin and paclitaxel. She died after 10 months due to septic shock [4].

The adjuvant chemotherapy regimen varies in reports (Table 1). Some authors used the bleomycin, etoposide, and cisplatin (BEP) regimen as in other germ cell tumors, and some used cisplatin,