Primary Extra Ovarian Dysgerminoma Associated with Endometriosis. Case Report

László Hodoniczki*, Alexandra Tóth, and Attila Keresztúri
Department of Obstetrics and Gynecology, University of Szeged, Hungary

Abstract

Background: To report a rare case of primary extra ovarian dysgerminoma associated with endometriosis.

Case presentation: A 34-year-old woman with known endometriosis had pelvic mass and underwent laparoscopy followed by laparotomy. The final pathological finding was primary extra ovarian dysgerminoma. There was a second operation. The patient got parametrectomy, omentectomy, right salpingectomy, sigma and ileum resection. The ovaries and the uterus remained free of disease. She got only one cycle of BEP chemotherapy, because she did not agree to participate in further chemotherapy. She gets now LhRH analogue therapy for endometriosis caused as cites, and she is in good condition.

Conclusion: The primary extra ovarian dysgerminoma is rare. There is no evidence on the treatment protocols, but surgery is essential.

ABBREVIATIONS

ESS: Endometrial Stromal Sarcoma; GnRH: Gonadotropin Releasing Hormone; LHRH: Luteinizing Hormone Releasing Hormone; FNAC: Fine Needle Aspiration Cytology; BEP: Bleomycin-Etoposide-Cisplatin based chemotherapy

INTRODUCTION

Endometriosis is an estrogen-dependent chronic common gynecological disorder in which endometrial tissue (glandular epithelium and stroma) is found at locations outside of the uterine cavity [1]. It affects 15% of women in childbearing age and 2-5% of postmenopausal women. Its incidence has been improving due to widely used laparoscopic operations. Although this disease is a benign condition, it has some malignant features, e.g. invasion of other tissues and organs, recurrence, appearance in distant locations. The etiology is not known, but the widely accepted mechanism is the adhesion and growth of endometrial fragments deposited in the peritoneal cavity via retrograde menstruation [2]. In 1925, Sampson was the first to document a case of suspected malignant transformation of endometriosis. Since then, several studies have focused on the relationship between endometriosis and gynecological cancer, especially ovarian cancer. Data from large studies indicate that endometriosis patients have an approximately 3-fold significantly higher risk of endometrioid and clear cell ovarian cancer. All types of malignant tumors were described in ectopic endometriosis. The most frequent tumors are endometrioid tumors and endometrial stromal sarcoma (ESS) [3].

Malignant ovarian germ cell tumors account for less than 5% of ovarian tumors. The peak incidence occurs in the mid and late teens. Dysgerminomas are the most frequently occurring malignant ovarian germ cell tumors. Primary extra genital dysgerminoma is rare and mostly occur in young man [4].

There is no data found in the literature of dysgerminoma and endometriosis.

CASE PRESENTATION

A 34-year-old woman presented with left lower abdominal pain and infertility. In the medical history there were two laparoscopic operations performed at age 30 and 31 due to chronic pelvic pain. Results of both operations were histological confirmed endometriosis. After the first operation the patient received GnRH analogue therapy for six months, following the second operation she remained asymptomatic for 2 years. Then abdominal pain came back. The ultrasound was negative. The patient received oral dienogest therapy for 6 months. After the therapy the pain improved, control ultrasound revealed an inhomogeneous mass of 25x30 mm size in the pouch of Douglas. There were no further investigations before the operation, like tumor markers and CT or MRI, because the patient has endometriosis in the history and it was thought, that this is a recurrence. We decided to perform a laparoscopy.
again. A bleeding tumor mass in the Douglas cavity was found. The operation was converted into laparotomy and removal of the tumor was performed. The intra-operative histopathology proposed hematologic malignancy, therefore the operation was finished. The final pathologic result confirmed dysgerminoma. The CT scan of the abdomen showed a heterogeneous mass in the region of the right ovary of 80 mm size. The uterus and the left ovary were normal, there was no lymphadenomegaly. The CT scan of the chest, the bone scintigraphy was without abnormalities. Tumor marker were done, the NSE and CA-125 were elevated, 16,7µg/l (normal range <16,3) and 72,57 U/ml (normal range <35,0) respectively, CEA was normal. The patient underwent a second operation. During surgery extensive adhesions were found. After dissection of the adhesions parametrectomy, omentectomy, right salpingectomy, sigma and ileum resection were performed. The ovaries and the uterus remained free of disease. The final pathologic result showed deep infiltrating endometriosis with extra genital dysgerminoma. The immune histopathology showed vimentin and PLAP positive, actin and CK negative tumour (Table 1).

Two years later, following the operation and one cycle adjuvant BEP chemotherapy the patient has worsening as cites during menstruation, caused by recurrent peritoneal endometriosis. She did not agree to participate in further chemotherapy. She is now receiving LHRH analogue therapy, she is in good physical condition, has amenorrhea, and has FNAC and cytologically documented recurrent disease in the abdominal wall. The last abdominal MRI showed some fluid in the abdominal cavity with no unequivocal tumorous lesion in the pelvis and abdomen.

### DISCUSSION

There is no specific marker for malignant transformation of endometriosis. Patients often have elevated blood CA125 concentrations, but normal HE-4 level. Imaging does not enable us to diagnose transformational forms. There are some suspect signs, eg fast-growing endometriomas with size more than 10 cm. Carcinoma is suggested in case of an exclusively solid mass or detection of papillary excrescences or focal solid areas. Diagnosing endometriosis-associated cancers can be difficult and it needs more immune histopathologic examinations. Immuno histochemistry is extremely valuable for determining the origin of the tumor [5]. Women most likely represent a different class of patients than traditional ovarian cancer patients and may require different therapeutic options. The surgical procedure and maximal cytoreduction is necessary. Further therapy depends on the correct histopathologic diagnosis. Some of the symptoms are caused by the endometriosis therefore we need to remember treating these separately from the tumor. Further investigation must focus on delineating the genetic, immune histochemical differences between women with endometriosis-associated extra-ovarian and endometriosis-associated ovarian cancer. [4-5]. There is no evidence on the treatment options, but the surgery is essential.

### REFERENCES


