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Case Reports

Rare case of coexistence of primary ovarian carcinoid in mature teratoma with primary serous carcinoma in second ovary – a case report

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Summary

Ovarian malignant tumours are mostly ovarian cancers. The most frequent ovarian benign lesions are mature teratomas. A very rare ovarian neoplasm is carcinoid. It mostly occurs as a component of mature teratoma, what causes rare diagnosis before surgery. Study presents the case of patient with primary ovarian carcinoid in mature teratoma of one ovary, co-existing with primary epithelial carcinoma of another ovary. Surgical treatment of carcinoid involves adnexectomy or hysterectomy with adnexa and removal of great omentum, followed by chemotherapy and radiotherapy. In young women with early-stage tumours, treatment can be limited to adnexectomy followed by close monitoring. In the presented case, management associated with the diagnosis of ovarian carcinoid, resulted in the detection of early-stage ovarian epithelial cancer. This case seems to confirm the recommendations to take tissue samples from the other ovary for histopathological evaluation in cases of ovarian unilateral benign tumours.

Key words: Carcinoid; Teratoma; Serous carcinoma.

Introduction

Ovarian malignant tumours, usually occurring in the peri- and postmenopausal periods, are mostly ovarian cancers. In Poland, ovarian cancer comes after breast, lungs, uterine corpus, and the large intestine cancers to become the fifth most common malignant tumour in women. It is believed to arise from the epithelial cells covering the ovary. The morbidity rate for ovarian cancer is 11.2/100 000 per year. In Poland, over 3,000 new cases are recorded each year. The risk of this tumour suddenly increases in the fifth decade of life, and successively raises up to the eighth decade. Typically, clinical symptoms occur late, when the disease is already advanced [1, 2]. Approximately 40-50% of ovarian cancers are serous tumours. Some of the most frequently diagnosed benign lesions of the ovary are mature teratomas. They constitute 45% of all ovarian neoplasms, including 58% of benign tumours. About 80% of mature teratomas develop in the reproductive age, especially in the second and third decades of life. Teratomas are rated among germ-cell neoplasms. Tissues that they consist of are mainly ectodermal derivatives, such as sebaceous and sweat glands, epidermidis, and hair follicles [3]. Malignant transformation takes place in one to three percent of cases.

A very rare ovarian neoplasm is a carcinoid, which constitutes about 0.3% of ovarian tumours. Primary ovarian carcinoid, which represents 0.5-1.7% of all types of carcinoid tumours, belongs to the group of neuroendocrine neoplasms. It mostly occurs as a component of mature teratoma, which is why it is rarely diagnosed before surgery. As a rule, the diagnosis is based on the histopathological evaluation of the surgically removed ovarian lesion. There are no randomised studies on this issue, and the knowledge of biology, prognosis, and treatment comes from retrospective and case studies. Consequently, there are no uniform guidelines for the management of the disease [4]. In the available literature, the authors have not found reports on the simultaneous co-existence of epithelial carcinoma of one ovary and carcinoid of another ovary.

This study presents the case of a 43-year-old patient with primary ovarian carcinoid arising in mature teratoma of one ovary, co-existing with primary epithelial carcinoma of another ovary.

Case Report

In March 2011 in the Department of Gynaecology and Urogynaecology, a 43-year-old patient was hospitalized for a focal lesion of the left ovary. The lesion was being monitored from 2007, when it was visualised in the ultrasound examination of the abdominal

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cavity. Last year, the patient complained of hot flush episodes. On admission, the subjective and objective examination, as well as laboratory tests did not show any significant deviations. Transvaginal ultrasound examination revealed: six-mm-thick homogeneous endometrium, a follicle with a diameter of 2.0 cm in the right ovary, and a cyst-like lesion with a diameter of 2.82 cm and image suggesting teratoma in the left ovary. The patient was qualified for laparoscopic enucleation of the cyst. During the surgery, a two-cm dermoid cyst was found in and totally enucleated from the left ovary. The surgery and the postoperative period were uneventful. The histopathological analysis of the material collected from the left ovary demonstrated a carcinoid tumour with a diameter of four mm (IHC: CHR+, SY 38+, CD 177-, vim, KL-1+, MIB-1<1% of cells) arising in mature teratoma. In accordance with the recommendations of the Oncological Committee, the patient was qualified for more radical surgery involving adnexectomy on the side of the lesion, appendectomy, obtaining tissue samples from the other ovary, and the removal of the greater omentum. The procedure was performed on April 6, 2011 in the Department of Gynaecology and Urogynaecology. The histopathological evaluation of the material taken during the surgery showed: focal resorption granulomas with foreign body giant multinuclear cells in the left ovary, the unchanged Fallopian tube, papillary serous carcinoma in the samples from the right ovary, numerous psammoma bodies (IHC: CK7, CK20, SY-38, CgA, CD 34, MIB-1), and unchanged omentum and appendix. The patient was re-consulted by the Oncological Committee, which recommended radical surgery and first-line chemotherapy. Hysterectomy and the right adnexectomy were performed in the Department of Gynaecology and Urogynaecology. The histopathological evaluation of the removed reproductive organs revealed infiltration from papillary serous carcinoma (G1) with psammoma bodies (the shortest distance from the outer surface of the ovary was 0.1 mm) in the right ovary. No neoplastic cells were found in the washings and swabs from abdominal cavity. The postoperative period was uneventful. Additionally, hormonal diagnosis of the patient was made in the Clinic of Endocrinology. During hospitalization no deviations from normal were detected in the objective examination. Laboratory tests demonstrated a slight increase in the cholesterol, ALT, and AFP levels. The ultrasound image of the thyroid gland suggested a chronic autoimmunological process, which was confirmed by the antibody tests against thyroid peroxidase. The patient was diagnosed as having Hashimoto's disease. The tests of the hormone levels (FSH, LH, PRL, PTH, cortisol, ACTH, insulin, and GH) did not show any changes. Also the levels of calcitonin, 5-hydroxyindoleacetic acid, beta-hCG, IGF-1, carcinoembryonic antigen (CEA), and chromogranin were within normal ranges.

After surgical treatment and endocrinological diagnosis, the patient underwent six-cycle chemotherapy. Follow-up examinations (computed tomography, tumour markers, indoleacetic acid, chromogranin) did not show any signs of a recurrent neoplastic process. At present, the patient is under the care of the endocrinology and oncology outpatient clinics (no complaints or deviations from normal during checkups). The recurrence of primary diseases has not been noted so far.

Discussion

This study presents the case of the premenopausal patient with a diagnosis of primary carcinoid arising in mature teratoma of one ovary and primary serous papillary carcinoma of another one.

Ovarian carcinoids belong to neuroendocrine neoplasms. They can occur as primary tumours, components of teratomas or mucous tumours (carcinomas, borderline tumours, benign cystic lesions), or secondary (metastatic) tumours. There are cases of lesions occurring in another ovary many years after surgical carcinoid treatment based on unilateral removal of the adnexa [5]. Most carcinoids arise in a dermoid cyst. Primary ovarian carcinoid was described for the first time in 1939 by Steward *et al.* [6]. In 1975 Godwin after the analysis of 2,837 cases of carcinoid tumours, gave details of three cases of primary ovarian carcinoid [7]. In 30% of cases, carcinoid causes typical symptoms of the carcinoid syndrome [8-10]. The cells of these tumours can produce substances such as: serotonin, bradykinin, histamine, the peptide YY, and gastrin. Hence, patients may complain of hot flushes, redness of the skin, diarrhea, stomach ache, and tachycardia. The carcinoid syndrome usually affects patients with the islet cell tumour [4, 11-14]. In the presented case, the patient only had hot flushes without any other symptoms of the carcinoid syndrome, therefore a diagnosis of this tumour before surgery was in her case practically impossible, especially because the diameter of a lesion was merely four mm.

A carcinoid component in a dermoid cyst is usually diagnosed postoperatively after histopathological evaluation [11, 15]. Carcinoids are mostly unilateral tumours, but it occur in 15% of cases that mature teratoma, mucous tumour, or Brenner's tumour occurs in the other ovary [16]. A vast number of carcinoids (almost 50%) are diagnosed in the early phase of development, which gives a good prognosis. In such cases, the five-year survival rate is 90%. [4, 12, 17]. This is true that they are rare neoplasms, and information about biology, prognosis, and treatment comes exclusively from retrospective studies. Consequently, it is difficult to establish uniform guidelines for the management of patients with such diagnoses [4].

Surgical treatment involves the resection of the uterine adnexa or the uterus with adnexa and great omentum, followed by chemotherapy and radiotherapy. Carcinoids are not particularly sensitive to chemo- and radiotherapy, but in cases of malignant tumours, surgical treatment should be combined with platinum and etoposide-based radiochemotherapy. In young women who want to preserve their fertility, and have early-stage tumours, treatment can be limited to adnexectomy followed by close monitoring. The co-existence of such tumours as mature teratoma and carcinoid is usually observed in one ovary [15]. Malignant transformation mainly occurs in patients above 40 years of age. Preoperative diagnosis of dermoid tumours is possible due to a quite characteristic ultrasound image. The treatment of mature teratomas involves enucleation of the whole tumour. It is acceptable to perform laparoscopic surgery with an endobag.

The most important prognostic factor in ovarian cancer is the stage of its development. Unfortunately, clinical symptoms of ovarian cancer manifest relatively late, there-

fore these tumours are usually diagnosed when they are already very advanced. Recommended methods of surgical management at the early stage of the disease are: abdominal hysterectomy with adnexectomy, the removal of the greater omentum, and appendectomy. Peritoneal washings and swabs should be collected. Next, patients are usually qualified for postoperative chemotherapy based on paclitaxel and the derivatives of platinum. In the presented case, further management associated with the diagnosis of ovarian carcinoid resulted in the detection of early-stage ovarian epithelial cancer.

The literature describes cases of the co-existence of histologically diverse tumours. This refers both to the presence of different tumours in the same ovary, and their presence in both ovaries. There is a number of reports on patients with carcinoids arising in teratoma tissue [15]. It occurs that ovarian cancers co-exist with the large cell neuroendocrine carcinoma (LCNEC), which belongs to primary ovarian neuroendocrine tumours. There is also the description of ovarian serous carcinoma cells and neuroendocrine carcinoma cells arising in one tumour [12, 13, 18-20]. The available literature does not provide data about the co-existence of primary ovarian cancer of one ovary, and carcinoid arising in teratoma of another ovary.

In the case presented above, it can be said that considering the small sizes of the tumours and the lack of clinical symptoms, the histopathological diagnosis, and the presence of neoplastic tissues in both ovaries came as a surprise. Fortunately, neoplastic disease was diagnosed early, which gave a good chance of complete recovery.

This case seems to confirm the correctness of the recommendations to obtain tissue samples from the other ovary for histopathological evaluation in cases of ovarian unilateral benign tumours.

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