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A case of delayed diagnosis of endometrial carcinoma due to a uterus didelphys

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Summary

In this case report, the authors present a patient with postmenopausal bleeding in whom diagnosis was delayed due to a uterus didelphys. Early diagnosis is important in endometrial carcinoma to ensure the highest possible chances of survival. In case of postmenopausal bleeding, physicians should be aware of the possibility of a uterus anomaly, especially in the presence of related congenital abnormalities as renal abnormalities. If there is any clinical suspicion of a uterus anomaly, such as uterus didelphys, further investigations such as MRI are needed and biopsies of both horns of the uterus are indicated.

Key words: Uterus anomaly; Müllerian duct anomaly; Uterus didelphys; Postmenopausal bleeding; Endometrial carcinoma.

Introduction

Endometrial cancer is worldwide the fifth most common malignancy in women with an estimated incidence of 320,000 new cases in 2012 worldwide [1]. Early diagnosis is very important, as early recognition and therapy have significant positive influence on morbidity and mortality. Five-year survival ranges from 74% to 91% in early stage disease (FIGO I or II), to 20–26% in late diagnosed carcinoma (FIGO IV) [2]. In case of a uterus anomaly, such as a uterus didelphys, diagnosis of a malignancy can be delayed, even when normal diagnostic protocols are followed. In this report the authors describe such a case in order to underline the importance of the critical eye of the clinician in cases presenting with postmenopausal bleeding.

Case Report

A 55-year-old woman presented with irregular postmenopausal bleeding and weight loss, five years after her menopause. She had no previous pregnancies. Her medical history revealed a lumpectomy because of breast cancer (T1cG2N0) and a congenital absence of the right kidney. Clinical examination of abdomen, vagina, and the cervix did not show any abnormal findings. Transvaginal ultrasonography showed an endometrial thickness of 13 mm and the shape of the uterus was slightly irregular. The ovaries were normal. The Pap smear was normal. Endometrial biopsy revealed inactive endometrial tissue, without any signs of hyperplasia or atypia. The results were discussed with the patient and she was advised to make a new appointment if the bleeding recurred. After eight months the patient returned with another episode of postmenopausal bleeding. This time the bleeding was severe. On physical examination a large subvaginal hematoma on the anterior wall was seen which had discharged spontaneously through an opening in the distal vagina. From here, profuse blood loss was observed. Transvaginal ultrasonography showed a possible heart-shaped uterus, with a clearly thickened endometrium. Because of profuse blood loss, it was decided to perform an examination under total anaesthesia with a gynaecologist specialized in oncology. The large hematoma of the anterior vaginal wall was opened, clots were evacuated, and a second cervix on the right side became visible. From this second cervix, profuse blood loss occurred. Hysteroscopy was therefore not possible. As the bleeding became life-threatening, it was decided to proceed with a laparotomy. After opening the abdomen through a mid-line incision, a uterus didelphys was found and the right horn was completely replaced by tumor (Figures 1 and 2). Multiple metastases (> 2 cm) were located on the diaphragm, omentum, and on large parts of the mesentery of the small bowel. The para-aortic lymph nodes were suspiciously enlarged. Frozen section examination of a peritoneal metastasis showed adenocarcinoma. Based on the clinical and operative findings, a metastasized endometrial carcinoma was suspected. Optimal debulking could not be achieved mainly because of the extensive involvement of the mesentery of the small bowel. Total abdominal hysterectomy with omentectomy was performed. Histologic examination showed a poorly differentiated endometrioid adenocarcinoma with multiple metastasis in both ovaries and omentum. The disease was consistent with FIGO Stage IV. Patient was discussed in the tumor board and four weeks postoperatively she started with chemotherapy (carboplatin i.v. and placlitaxel i.v.). She died of progressive disease one year after the diagnosis.

Discussion

Congenital uterine malformations are due to a defect in formation, fusion or septal absorption of Müllerian ducts.[3] The incidence of Müllerian duct anomalies in a general population is approximately 5.5% to 9.8% [4, 5].

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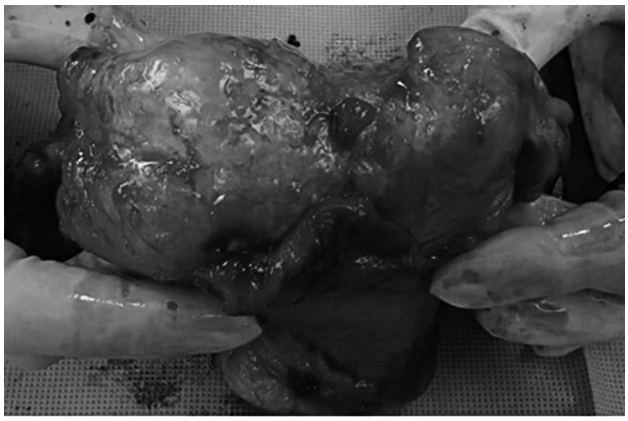

Figure 1. — Both horns of a uterus didelphys; the right horn is completely replaced by tumor.

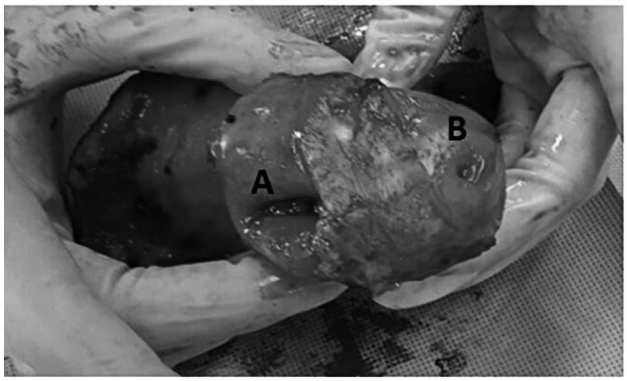

Figure 2. — A) Right ostium uteri. B) Left ostium uteri.

For uterus didelphys the estimated incidence is about 0.4% [6]. In woman with Müllerian defects, the reported incidence of associated renal anomalies is 20% to 30% [7, 8]. In the 1950s, the first case reports of a uterus didelphys in combination with an endometrial carcinoma were published [9-11]. So far, uterine malformations have not been associated with an increased risk for endometrial carcinoma [12]. Nevertheless, it is important to include the possibility of a uterus anomaly in the management of patients with postmenopausal bleeding. Diagnosis may be delayed if in case of a double uterus, only one horn would be biopsied. Additional imaging technique may be of use if a uterus anomaly is suspected, especially MRI, which is has a high sensitivity and specificity [13, 14].

In the presented case, the diagnosis of a uterine anomaly was missed. The unclear shape of the uterus at transvaginal sonography together with a history of a renal anomaly however, were sufficient reasons for further investigation such a MRI. Given the fact that the woman presented with advanced disease eight months later, it is likely that endometrial carcinoma was the cause of the first episode of postmenopausal bleeding.

In this case the endometrial carcinoma arose probably in one horn. One explanation for this might be that it was a grade 3 endometrioid carcinoma. There is growing evidence that this type of tumor shares similar clinical, histological, and prognostic features with type 2 endometrial carcinomas [15]. Type 2 tumors are non-estrogen dependant and can arise unifocally in an atrophic endometrium.

Treatment and prognosis between endometrial carcinoma arising in a normal uterus and in a uterus didelphys do not differ. Treatment and prognosis are mainly determined by FIGO stage and histologic type.

Conclusion

This case demonstrates that diagnosis of endometrial carcinoma can be a delayed in the presence of a uterus anomaly. Consequences may be detrimental as prognosis is significantly determined by the extent of the disease. In patients with postmenopausal bleeding, clinicians must be aware of the possibility of a uterus anomaly, and if necessary perform additional investigations such as MRI, in order to obtain reliable histological endometrial sampling.

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