Adenosarcoma of the uterine body in a 19-year-old woman - three year survival: Case report

T. Fait, *M.D.*; **J. Zivny,** *M.D., Ph.D.*; **P. Freitag,** *M.D., Ph.D.*; **D. Kuzel,** *M.D., Ph.D.*

Department of Gynecology and Obstetrics, 1st Faculty of Medicine, Charles University, Prague (Czech Republic)

Summary

Background: Uterine adenosarcoma is a rarely by occurring tumor. It is composed of a benign adenoid structure and a sarcomatous stromal component. The average age of patients with a diagnosis of uterine adenosarcoma is about 70 years.

Case: We present a case of a 19-year-old woman with a rarely occurring uterine adenosarcoma manifesting itself by irregular bleeding and producing fragile polypous matter which was spreading into the vagina. The final diagnosis was made only by repeated biopsies. Abdominal hysterectomy with bilateral salpingo-oophorectomy, appendectomy and revision of iliac lymph nodes were performed. Teleradiotherapy was applied from 4 fields in 25 fractions to a total exposure of 50 Gy. It was followed by six cycles of chemotherapy containing 50 mg/m^2 doxorubicin and 5g/m^2 ifosfamid administered in 21-day dose intervals.

Conclusion: This case should demonstrate the difficulty of making the right diagnosis. Since the end of therapy the patient has been regularly seen in our onco-gynecologic department. Now, 40 months after the end of chemotherapy and 46 months after making the diagnosis, there are no signs of relapse.

Key words: Adenosarcoma uteri; Teenager; Case report.

Introduction

We present a case of a 19-year-old woman with a rarely occurring uterine adenosarcoma manifesting itself by irregular bleeding and producing fragile polypous matter which was spreading into the vagina. The final diagnosis was made only by repeated biopsies. This case demonstrates the difficulty of making the right diagnosis of such a rare oncological disease.

Case report

H.S. was born in August 1977.

Her personal and family histories were irrelevant. Her menstrual cycle was regular from the age of 15 till the age of 17. She was treated in another hospital for irregular metrorrhagia in November 1994 (age 17) and in February 1995. The metrorrhagia was cured by uterotonics, antifibrinolytics and progestins. Since March 1995, she has been taking monophasic contraception containing 125 µg levonorgestrel together with 50 µg ethinyl estradiol. As a result of using this medication, her cycle became regular. There was constantly a low level of LH (11.3-13.9 IU/l) and a higher level of testosterone (0.99 mg/l) in her hormonal profile in March 1996.

The patient was sent to our department for the first time in April 1996 with a diagnosis of myoma nascens and metrorrhagia. Her last menstrual period was regular and she was not febrile. Dark tissue protruded from the cervix with bad-smelling dark blood. The uterus was enlarged to the size of the 8th week of pregnancy. The level of HCG was repeatedly negative. Ultrasonic examination showed a straightened uterus with a dilated cavity and a highly nonhomogenous endometrium; both ovaries were slightly cystic. A great amount of necrotic tissue was obtained by instrumental revision of the uterine cavity. Histology revealed inflamed cellular necrotic tissue. Neither tumorous growth nor pregnancy was detected. The final diagnosis was dysfunctional metrorrhagia.

A regular menstrual cycle was maintained by medroxyprogesterone acetate, 5 mg per day, from the 15th to the 25th day of the cycle.

In July 1996, she was admitted to the hospital because of heavy bleeding during menses lasting ten days. There was a lot of coagulum in the vagina and a necrotic rounded formation 80 mm in diameter protruded from the cervix and filled the vaginal vaults. The cervix was rigid and dilated. The HCG test was negative. Norethisteron was applied to stop the bleeding and to prepare the uterus for hysteroscopy. Hysteroscopy findings were wide peduncle growing from the back wall carrying a polypous formation, soft and thin walls, and an uneven back wall. Histology a showed polypous formation with fuso-cellular edematous soaking stroma and glandular formations.

The histopathologist hesitated between two diagnoses: adenomyoma or endometrial stromal node. A second-look hysteroscopy with an examination after three months was recommended to the patient.

In the beginning of August 1996, H.S. was repeatedly hospitalized for heavy bleeding during menstruation. The vagina was full of clots. The cervix of the straightened uterus was permeable upto a finger. Ultrasound findings showed nonhomogenous dilatation of the uterine cavity. She was treated with uterotonics and hemostyptics. In preparation for the second-look hysteroscopy, norethisteron was administered.

She was again hospitalized at the end of August 1996 with strong metrorrhagia; she bled by clots, suffered from pressure in the hypogastrium and was subfebrile. The vagina was full of coagulum and dark blood, the cervix was cleft, and fragile tissue protruded. The uterus was enlarged to the size of the 11th week of pregnancy. Ultrasonic examination showed a cavity filled with solid hemorrhaged tissue (RI 0.35). Second-look hysteroscopy and curettage were performed. The pathologist obtained necrotic polypous matter similar to the remains of a

Revised manuscript accepted for publication October 5, 2000

miscarriage. Structures of regressively changed decidua and fragments of tissue that were supposed to be postmiscarriage residua were found. Spasmodic stripes of cartilaginous tissue and sheaves of striped muscular cells were found in edematous fuso-cellular tissue (Figure 1).

Two weeks later a third-look hysteroscopy with resection of part of the myometrium was done because of the diagnostic hesitancy. After reviewing all the materials, a diagnosis of adenosarcoma (Figure 2) of the uterus was made. The cartilaginous tissue and muscular parts were identified as tumorous and the presence of decidua was explained by the hormonal therapy.

Before the operation a cystoscopy and a chest X-ray had been performed with normal results, and computer tomography examination found an enlarged nonhomogenous uterus and a polycystic left ovary. Tumor markers (CA 125, CA 15-3, CEA, and CA 19-9) were negative.

At the end of September 1996, abdominal hysterectomy with bilateral salpingo-oophorectomy, appendectomy and revision of the iliac lymph nodes were performed. There was a polypous formation on the left sacrouterine ligament which was exsied. Peritoneal lavage cytology was examined with a negative outcome. Histology revealed the uterine cavity to be inlayed with grayish-white sharply bordered tumorous tissue 8 mm thick with polypous formations at the top of the cavity. The adenosarcoma was beginning to infiltrate the myometrium at multiple sites. The fuso-cellular component contained myxoid parts, islands of well-differentiated cartilage and tumorous rhabdomyoblasts. Metastasis on the peritoneal cover of the Douglas cavity was formed by a well-differentiated chondrosarcoma and myxoid tissue with fuso-cellular (spindle-shaped) malignant elements.

The postoperative course was without complications.

In October and November 1996 teleradiotherapy was applied from 4 fields in 25 fractions to the total exposure of 50 Gy. It was followed by six cycles of chemotherapy containing 50 mg/m^2 doxorubicin and 5g/m^2 ifosfamid, administered in 21-day dose intervals.

The patient reacted with a complete alopecia. Depression of haematopoesis with leukopenia required the application of prednisone. Cystoscopy after the sixth cycle demonstrated a normal picture.

In April 1997, after the end of chemotherapy, a control computer tomography examination was performed with an adequate postoperative finding. A normal structure of the parenchymatous organs was found. No focal changes were found by pulmonary radiography. Second-look laparoscopy had a normal

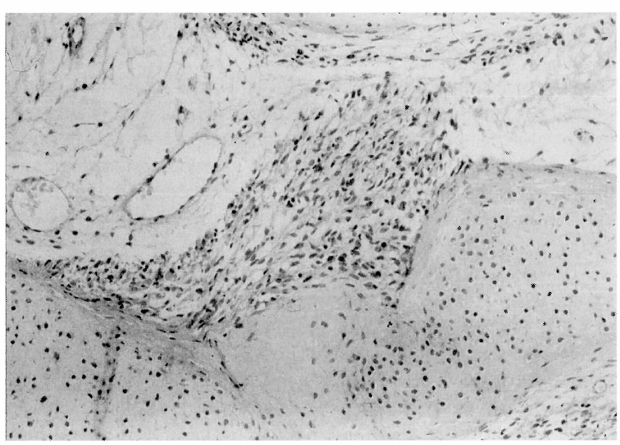

Figure 1. — The cartilaginous and striped muscular tissues in stromal part of the adenosarcoma.

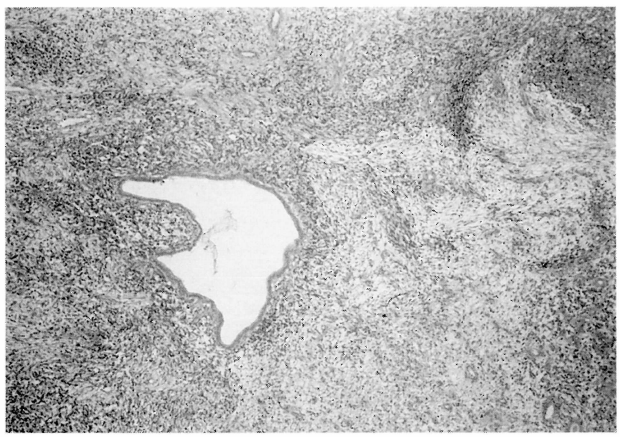

Figure 2. — Detail of adenosarcoma formed by benign glandula and malignant stromal part.

postoperative status. Lavage cytology of the abdominal cavity and bioptic samples from the cicatrix of the left sacrouterine ligament and the right suspended ovary ligament did not contain any tumorous elements.

Since then the patient has been regularly seen in our oncogynecologic department. Because of 80% positivity of estrogen receptors in the tumor, HRT has not been recommended. Bone density is at normal levels (Lunar Achilles). Now (August 2000), 40 months after the end of chemotherapy and 46 months after making the diagnosis, there are no signs of relapse.

Discussion

Malignant tumors of the uterine body form about 6% of tumorous disease in women [1]. Sarcomas, which represent about 2-7%, are divided into leiomyosarcoma, malignant mixed mesodermal (Müllerian) tumors and endometrial stromal sarcoma [2]. Clement and Scully [3] described adenosarcoma uteri for the first time in 1974. This tumor type is composed of two intermixed neoplastic tissues with a benign epithelial component and a sarcomatous stromal component.

The average age of Bloom's [4] patients was 76 years. Among patients of our oncology department we followed up a series of 27 women with gynecologic sarcomas over 20 years. Twenty cases were sarcoma of the uterus. The youngest patient was 42 years old which was a case of leiomyosarcoma. Müllerian mixed mesodermal sarcomas appeared three times; they were carcinosarcomas of postmenopausal women aged from 65 to 79. More than half of our series were patients with leiomyosarcomas. In a similar series of Jereczek [5], only one-third were leiomyosarcomas.

Marabini *et al.* [6] diagnosed a 40 mm long polypus composed of a stromal sarcoma in a 26-year old patient with recurrent metrorrhagia . Michalas *et al.* [7] found in a case of a 16-year old patient stromal sarcoma with primary focus in the rudimentary corn of the uterus. A clinical picture of an abdominal emergency showed dissemination into the lungs and the abdominal cavity.

Embryonal rhabdomyosarcoma (sarcoma Bothryoides)

is a tumor of the vagina or cervix of the uterus in young girls with maximal prevalence in the 2nd year of age [8].

In cases of women at the beginning of their reproductive age only endometrial stromal sarcomas have been diagnosed [9]. Other types of sarcomas have not been described so far in this age group of women.

The finding of spasmodic stripes of cartilaginous tissue and sheaves of striped muscular cells was the most difficult point of our case. It was wrongly determined as the remains of a miscarriage. It created diagnostic hesitancy because of repeatedly negative levels of hCG. In the beginning the heterologous type of adenosarcoma with well-differentiated chondrosarcoma was not taken into account.

References

[1] Chaplain G., Guisenier J., Milan C., Janoray P., Collins F., Dechelotte D., Michiels-Marrais D. *et al.*: "Cancers du corps de l'uterus dans le department de la Cote-d'Or 1982-90". *Bull. Cancer*, Paris, Aug. 1994, *81* (8), 691.

[2] Bloch B., Dehaeck R., Soters R.: "Manual of Practical Gynecological Oncology". Chapman and Hill, London, 1995, 198.

[3] Clement P. B., Scully R. E.: "Mullerian adenosarcoma of the uterus". *Hum. Pathol.*, 1990, *21*, 363.

[4] Bloom R., Guerrieri C.: "Adenosarcoma of the uterus". *Int. J. Gynecol. Cancer*, 1999, *9*, 37.

[5] Jereczek B., Jassem J., Kobierska A.: "Sarcoma of the uterus". *Arch. Gynecol. Obstet.*, 1996, *258* (4), 171.

[6] Marabini A., Gubbini G., De Jaco P., Stagnozzi R., Santini D.: "A case of suspected endometrial stromal sarcoma removed by operative hysteroscopy". *Gynecol. Oncol.*, Dec. 1995, *59* (3), 409.

[7] Michalas S., Greatsas G., Deligeorogiou E., Markaki S.: "High grade endometrial stromal sarcoma in a 16-year-old girl". *Gynecol. Oncol.*, Aug. 1994, *54* (1), 95.

[8] Dewhurst J.: "Practical pediatric and adolescent gynecology". New York, Marcel Dekker, Inc. 1980, 272.

[9] Sanyal C.: "Stromal cell sarcoma of uterus in a teenage girl". *J. Indian Med. Assoc.*, Nov. 1994, *92* (11), 391.

Address reprint requests to:
T. FAIT, M.D.
Department of Gynecology and Obstetrics
General Faculty Hospital
Apolinarska 18
Prague 2
12851 Czech Republic