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Pleomorphic rhabdomyosarcoma of the uterine corpus – a case report

Opis przypadku mięsaka prążkowanokomórkowego pleomorficznego trzonu macicy

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Abstract

Pleomorphic rhabdomyosarcoma of the uterus is a rare malignant tumor. It is connected with postmenopausal abnormal vaginal bleeding and abdominal pain.

We report a case of a 66-year-old postmenopausal woman diagnosed with abnormal vaginal bleeding and abdominal pain. Vaginal ultrasonography showed enlarged uterus, 82mm x 64mm in size. The patient underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy with postoperative chemotherapy due to pleomorphic rhabdomyosarcoma of the uterus. The patient died 2,5 years after the surgery as a result of a rapid spread of the neoplastic process. The case of rhabdomyosarcoma, together with the review of the literature, is presented in the following work. We find that the rarity of this histological entity makes it particularly worthy of publication.

Key words: **pleomorphic** / **rhabdomyosarcoma** / **uterus** / **adult** / **postmenopausal** / / **bleeding** / **sarcoma** /

Streszczenie

Mięsak pleomorficzny, prążkowanokomórkowy jest rzadkim nienabłonkowym nowotworem złośliwym mięśnia macicy, występującym u pomenopauzalnych pacjentek, które zgłaszają w wywiadzie nieprawidłowe krwawienia maciczne i dolegliwości bólowe podbrzusza.

W opisywanej sytuacji klinicznej 66 letnia wieloródka była hospitalizowana z powodu nieprawidłowego krwawienia z dróg rodnych oraz bólu podbrzusza. W badaniu USG uwidoczniono powiększony trzon macicy o wymiarach 82x64 mm. Przeprowadzono zabieg usunięcia macicy z przydatkami w sposób typowy. W badaniu histopatologicznym, potwierdzonym immunohistochemiczne, stwierdzono postać pleomorficzną mięsaka trzonu macicy. Po operacji pacjentka przeszła chemioterapię. Z powodu przerzutów pacjentka zmarła po około 2,5 roku od leczenia chirurgicznego. Z powodu bardzo rzadkiego występowania postaci pleomorficznej mięsaka prążkowanokomórkowego macicy, ten opis przypadku, łącznie z przeglądem literatury, wart jest opublikowania.

Słowa kluczowe: **postać pleomorficzna** / **mięsak prążkowanokomórkowy** / **macica** / / **menopauza,** / **krwawienie** / **mięsak** / **dorośli** /

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Introduction

Sarcomas of the uterine corpus are rare, non-epithelial malignant tumors which constitute 2% to 5% of all malignant uterine tumors [1]. Incidence rate is from 0,5 to 2/100 000 of women [2]. The most common types are: mixed mesodermal tumors, leiomyosarcomas, and endometrial stromal tumors. Other histological types, including e.g. rhabdomyosarcomas, liposarcomas, chondrosarcomas have very low incidence [3]. Rhabdomyosarcoma is a tumor with a very high degree of histological malignancy and is extremely rare in the uterine corpus. Three general pathologic types of rhabdomyosarcoma: embryonal, alveolar, and pleomorphic, have been described in literature [3,4]. Unfortunately, uterine pleomorphic rhabdomyosarcomas are rare and highly malignant tumors, with frequent extrauterine spread at presentation which makes it difficult to study the cases. Patients rarely survive longer than 15 months [5].

We present a case of a 66-year-old woman with postmenopausal vaginal bleeding and abdominal pain caused by pleomorphic rhabdomyosarcoma of the corpus uteri.

Case report

A 66-year-old multipara was admitted to clinical hospital of Poznań University of Medical Sciences in 2004 due to abnormal vaginal bleeding and abdominal pain.

Gynecological examination showed anteflexed and enlarged uterine corpus. Ultrasonography revealed uterine corpus to be 82mm x 64mm in dimension and 52mm thick, with fluid-filled endometrium constituting 2/3 of the uterine wall. In the interview the patient reported last menstruation at the age of 48 and a few years of hypertension therapy with Enarenal (10mg twice a day), Staveran and Lorafen (80mg and 2.5mg once a day, respectively). Obesity (BMI=39.5), gout and condition after cholecystectomy, performed in 1983 due to cholecystolithiasis, were found as well. Chest RTG showed aorta atherosclerosis. Dilation and curettage of the uterus were proposed due to clinical state and after gynecological examination. However, when the patient was under general anesthesia, the dilatation of external orifice of the uterus proved to be impossible. The procedure of curettage of the uterus cavity was aborted and hysterectomy with adnexectomy was performed instead. Laparotomy disclosed enlarged soft uterus, 6 x 7cm in dimension, which was then removed together with adnexa in a typical manner. In palpation examination of the liver, greater omentum and parietal peritoneum of pelvis minor no metastases were found. The patient was discharged home in overall good condition, without any ailments.

The result of the histopathological examination obtained after 14 days stated: 'Rhabdomyosarcoma pleomorphicum corporis uteri partim necroticum at haemorrhagicum. Endometrium atrophicum. Leiomyomata parva corporis uteri. Endocervicitis chronica. Erosio glandularis in statu epidermisationis. Fibrosis adnexorum'. Immunohistochemical analyses (Myf 4+, actin+, desmin+, vimentin+) that confirmed the diagnosis of pleomorphic rhabdomyosarcoma were carried out. Six cycles of postoperative chemotherapy (with 500 mg/m^2 intravenous (IV) Cyclophosphamide/1day, 1mg/m^2 IV Vincristine/1 and 5 day, 50mg/m^2 IV Doxorubicin/1day, 250mg/m^2 IV Dacarbazine 1 and 5 day) were given according to protocol. Follow-up examinations one year after surgery revealed no abnormalities or tumor recurrence. Eighteen months after the surgery FUSION PET (PET/CT) of facial skeleton, neck, chest, abdominal cavity and pelvis was performed and no foci of increased glucose metabolism corresponding to active proliferative process were observed. Unfortunately, the patient died 2.5 years after the surgery due to rapid spread of neoplastic, intraperitoneal process and metastases to the lung and liver.

Discussion

Sarcomas account for <5% of endometrial curettage. The most common types, over 95% of all uterine sarcomas, include mixed mesodermal tumors, leiomyosarcomas and endometrial stromal tumors. Other histological types, e.g. rhabdomyosarcomas, liposarcomas and chondrosarcomas, have very low incidence (0.05/100 000 women) [3]. Three general pathologic types of rhabdomyosarcoma, i.e. embryonal, alveolar, and pleomorphic, have been described in literature [4]. Embryonal and alveolar rhabdomyosarcomas occur in girls and young women, whereas pleomorphic rhabdomyosarcoma is diagnosed in elderly, postmenopausal patients, usually above age 45 [2, 3]. Pleomorphic rhabdomyosarcoma has very high degree of histological malignancy and is extremely rare in the uterine corpus.

Risk factors are similar to those of endometrial carcinoma. The most frequent symptom, reported by 80% of patients, is abnormal vaginal bleeding [2, 3]. The bleeding is accompanied by abdominal pain resulting from uterine contractions and removal of fragments of neoplastic tissues from the genital tract [2,3]. Gynecological examinations reveal enlarged uterus and palpable tumor in pelvis minor in 20-40% of patients [3]. Ultrasound diagnostic of endometrial thickness is also usefulness tool in diagnosis of endometrium pathology in patients with abnormal postmenopausal vaginal bleeding [6]. In the described clinical case the 66-year-old patient was hospitalized due to abnormal postmenopausal vaginal bleeding and abdominal pain, i.e. symptoms of low specificity.

Histopathological analysis of the material from curettage of the uterus is the basis of preliminary diagnosis of neoplastic changes. Final diagnosis of the degree and extent of neoplastic process is based on the results of histopathological analysis of the tissue sampled during operation (among others, preparation of the uterus with adnexa and greater omentum and lymph nodes biopsy). In case of our patient, the dilatation of external orifice of the uterus was impossible and the procedure of curettage of the uterine cavity was aborted. The surgery included only hysterectomy with adnexectomy as the pleomorphic rhabdomyosarcoma of uterine corpus was diagnosed only after histopathological analysis of operational material and immunohistochemical examination confirming the diagnosis were performed.

Open-access medical data bases include few cases of pleomorphic rhabdomyosarcomas of the uterine corpus. Observations of Norddal and Thoresen [7] carried out from 1956 to 1992 in a group of 1042 women diagnosed with and treated for sarcomas of the uterine corpus, showed that only 11 patients suffered from pleomorphic rhabdomyosarcoma [7]. Similarly, in studies lasting 27 years (since 1974 to 2001) conducted by Livi et al. in a group of 141 women only one case of rhabdomyosarcoma was confirmed in histopathological examination [8]. Hart et al. described two botryoid embryonal and two pleomorphic rhabdomyosarcomas in adult patients [9].

Both patients, with typical botryoid embryonal rhabdomyosarcomas were alive and well, following combined treatments with surgery and chemotherapy or irradiation, 3.0 and 2.7 years after the operation. The other two women, with pleomorphic rhabdomyosarcomas, died rapidly of widespread tumor [9]. In another study Borka et al. described a case of a 67-year-old woman with postmenopausal vaginal bleeding caused by pleomorphic rhabdomyosarcoma of the uterus, treated with laparotomy and postoperative chemotherapy [10].

Pleomorphic rhabdomyosarcoma of the uterine corpus is a sarcoma of high degree of histological malignancy. It consists of multiform, sometimes fusiform, big and often large multinuclear neoplastic cells with acidophilic cytoplasm. Neoplastic cells are chaotically distributed in connective tissue stroma and the transverse striation is almost never observed [2]. Immunohistochemical analysis is an essential tool verifying histological types of rhabdomyosarcomas. However, expression of desmin and muscle actin is noticeable [2].

Treatment of uterine sarcoma relies on precise diagnosis of its histological form and clinical evaluation of its progression. Resulting chemotherapy is based mostly on antracyclins and ifosfamide [11]. Adjuvant chemotherapy with VAC (vincristin, actinomycin D and cyclophosphamide) may also improve survival [12]. Yeasmin et al described a case of a 60-year-old postmenopausal woman diagnosed with pleomorphic rhabdomyosarcoma. After surgery and systemic chemotherapy and pelvic irradiation with a high lactate dehydrogenase (LDH) level, the patient died within 20 months of the diagnosis [13]. Borka et al published a case of a 67-year-old woman who underwent laparotomy and postoperative chemotherapy to survive one year after the surgery [10]. While radiotherapy reduces the frequency of recurrence, its impact on a 5-year survival has not been proven [2]. However, there is evidence that patients with embryonal and alveolar rhabdomyosarcomas live long enough for the effect of multicomponent therapy on occurrence of other tumors to be accessible. These are usually osteosarcoma (caused by radiotherapy) or non-lymphatic leukemia (complication of chemotherapy) and the risk of their appearance is 1.7% in the course of 10 years [2]. It seems that combined therapy (surgery with chemo and radiotherapy) has resulted in long-term survival for majority of patients with urogenital rhabdomyosarcomas [10].

Pleomorphic variant rhabdomyosarcomas are very aggressive neoplasms with extremely poor prognosis. Almost 2-year survival of our patient after surgery with no detection of any recurrences may be considered a success.

Conclusions

Summing up, the above-described clinical case of pleomorphic rhabdomyosarcoma of the uterine corpus is a very rare malignant tumor. Its diagnosis is based on histopathological analysis confirmed by immunohistochemical examination and clinical symptoms are non-specific. The rarity of this histological entity and protocol applied make the presented case worthy of publication.

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