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Kaposi's sarcoma of unusual localization

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Abstract

27 years old patient admitted to our unit for surgical treatment due to AUB with HCV infection patient was qualified for aputation of the corpus of the uterus. Procedure was performed without any complications. During hospitalisation HIV virus infection was found. In histopathological examination of corpus of the uterus Kapossi sarcoma was found. Authors were unable to find any description of such localization of Kapossi sarcoma.

Keywords: *Kapossi sarcoma, uterus, AUB*

A case report

A 27-year-old female patient of NR was admitted to the Gynecology ward for a scheduled hysterectomy without the adnexa. The indication for surgical treatment was recurrent menorrhagia causing significant anemia, requiring treatment with iron preparations. Attempted conservative management through the use patient underwent cholecystectomy 7 years ago. One year after surgery, she was diagnosed with viral hepatitis C. She was treated with interferon. During the admission to the Gynecology Department, symptoms of steatosis and elevated transaminases were observed.

As a response to the patients' complicated situation and the lack of treatment efficiency with hemorrhages, surgical treatment was started and curettage of the uterine cavity was performed several times, but only with short-term effects.

Histopathological examinations of the obtained material revealed endometrial hyperplasia. In the final test, the histopathologist found atypical cells in the endometrium. After the patient's general state of health, the fact that she has three children and does not plan another, the patient was offered a hysterectomy without appendages, for which she has agreed.

The operation was performed without complications, but during it, an accidental needle stick occurred to one of the members of the operating team.

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The patient was immediately subjected to virological tests, which, to the surprise of the medical team, showed that the patient was infected with HCV and HIV. The result was confirmed with the Western Blot test. Post-exposure prophylaxis was applied by a doctor. The tests carried out on him after 3 weeks gave a negative result.
The virological examination was recommended by the patient's partner. The histopathologist, who did not know the result of the virological examination, after assessing the surgical specimen, found out if the patient was not infected with HIV because he diagnosed Kaposi's sarcoma with such an unusual localization.

Results of the examination:
Material for examination was obtained from the uterine corpus of 80,0 g and dimensions 6,5x6,0x3,0cm.

The material was examined serially by making numerous microscopic preparations. Uterine wall in the endometrium of the corpus, multifocal lemma of blood vessels arranged in a nodular structure. The histological picture resembles a hemangioma with scanty inflammatory infiltrates on the outside of the intraepithelial cells and fibroblast-like cells which form distinct nodules in several places
Neogensis of proliferating vessels is also clearly visible in the myometrium

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Neogensis of proliferating vessels is also clearly visible in the myometriumThe expression of factor VIII, CD31 And CD34 was observed in the intravascular space of proliferating nodules but not in Fibroblast-like cells. Increase in the activity of proliferating factor Ki 67 in the intravascular membranes.
The histological picture based on the findings of the immunohistochemical report and clinical data corresponds to the second stage (hemangioma) and the third stage (nodular) form of Kapossi sarcoma.

The tumor, known today as Kaposi's sarcoma / KS /, was described in 1872 by the Hungarian dermatologist Moritz Kaposi and called it idiopathic disseminated pigmented skin sarcoma. Kaposi's sarcoma is a soft tissue neoplasm originating in the endothelium of the lymphatic vessels. It rarely metastasizes / 1 / and often remains asymptomatic for many years. It comes in 4 clinical varieties:
1.Classic 2. Epidemic associated with AIDS 3. Endemic African 4. Iatrogenic after immunosuppression.
KS usually affects the skin, rarely internal organs / 2 /.
All clinical variants of KS are caused by one or more of the six subtypes A, B, C, D, E, F (3,4,5,6,7) of the HHV-8 herpes virus, termed. by Chang et al as Kaposi sarcoma-associated virus KSHV / 8 /. In 70-100% of patients, KSHV is found in tumor cells. / 9 /. The HHV - 8 virus cofactor in the development of KS is the secondary decrease in immunity in HIV infection and AIDS patients or a radical decrease in immunity after organ and tissue transplants due to the use of immunosuppressive drugs. / 10 /. Horizontal transmission of the HHV-8 virus takes place mainly through saliva / 11, 12, 13, 14, 15, 16 /. HHV-8 is found in the saliva of 61% of infected, a lower percentage in semen and vaginal secretions, less frequently than HBV and HCV, it is transmitted during intravenous drug administration and needle- sharing. KS usually develops 15-30 months after infection with KSHV.

The most aggressive and rapidly progressive form of KS, also affecting internal organs, occurs in patients with AIDS. KS is the so-called indicator disease determining the diagnosis of full-blown AIDS / clinical category C /. / 2,17 /
The wide clinical and pathological spectrum of KS suggests that it is not a single disease. The predictive value of the clinical diagnosis of KS is low / 18 /. Histopathological confirmation of the diagnosis remains the gold standard, but it is difficult, especially for a pathologist without experience in the assessment of a wide spectrum of KS morphology images.
Treatment of KS, due to the multifaceted nature of the disease, the degree of advancement and the location of the lesions, as well as etiological differences related to multiple KSHV subtypes, requires the use of various methods. These include surgery, laser, cryotherapy, photodynamic method, radiation therapy, chemotherapy, antiviral drugs, interferon. Antiretroviral therapy is also effective in the early stages of KS.

Discussion

Kaposi's sarcoma from the date of its description to the 1980s was a rare disease of unknown etiology. In its classical form, it was found in the Mediterranean basin, mainly in men over 50, most often in Ashkenazi Jews / 2,20,21 /. In the endemic variety, it was found in Central Africa in young men and children / 2 /. The incidence of KS has increased rapidly since the beginning of the 1980s, due to the development of the HIV pandemic and the spread of immunosuppressive drugs related to the development of transplantology. Numerous scientific studies were undertaken in those years that allowed to elucidate the etiology of the disease and linking it with the herpes virus HHV-8 and immunodeficiency.

Only the introduction of effective antiretroviral therapy in the treatment of AIDS in the mid-1990s resulted in a reduction in the number of new cases by 60–70% / 22 /. Kaposi's sarcoma develops mainly in the skin and subcutaneous tissue. The extra-cutaneous form associated with the aggressive epidemic variant of KS accompanying AIDS is observed less frequently. It includes neoplastic changes in the liver, lungs, gastrointestinal tract, lymph nodes, and the most common nowadays, in the oral mucosa. KS was very rarely featured elsewhere.
There is no description of a case of uterine sarcoma in the world literature. This location is very difficult to detect and the symptoms of the disease are unusual. Menorrhagia periods have many causes. Women constitute only 6% of patients with KS, as the vast majority of patients are homosexual men / 23 /. The therapeutic curettage of the uterine cavity, combined with histopathological examinations, performed several times in our patient, did not indicate the presence of neoplasm. We cannot explain this fact. Perhaps the neoplastic lesion was located in the deep layers of the endometrium on the border of the uterine muscle and was not found in the tissues obtained during curettage. We admire the histopathologist who did not have relevant data and did not have much experience in diagnosing KS, yet made a difficult diagnosis correctly.
Retrospectively, we consider it a mistake or oversight that a patient treated by an infectious disease specialist for hepatitis C was not tested for HIV. This test is nowadays a diagnostic standard.

Conclusions

The described case introduces into oncology a new, not previously described location of Kaposi's sarcoma and confirms the statement that any location of this tumor is possible. The practical conclusion is to sensitize doctors to the need to test for HIV infection in all patients with diseases associated with the risk of coexisting this infection.

Histopathological examination

Material given under the histopathological examination consists of 80,0g uterine corpus and size 6,5x6,0x3,0cm.

The material was investigated with many microscopical slices.

Uterine corpus.

In the endometrium extensive vascularisation, taking a shape of a tumor. The histological aspect shows hemangiomas with minor inflammation. Outside of the endometrial cells, we can observe long fibroblast-like cells, which in few places show a distinguished tumor.

Proliferating vasculature is formed in the uterine muscles.

The immunohistochemical examination has shown the expression of factor VIII, CD31, and CD34 in the endothelium of progressing tumors but not in the fibroblast - like cells. Increased activity of Ki

)7 has been observed in the endothelium.

Histopathological examination based on described previously observations made is consistent with second (vascular type) and third stage (tumor-like) Kaposi sarcoma.

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