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Uterine corpus rhabdomyosarcoma in 13-year-old girl

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Embryonal rhabdomyosarcoma (RMS) is the most common malignant tumor of the genitourinary system in children. The most common symptoms in girls include vaginal bleeding, abdominal pain, urinary and/or stool incontinence and hematuria. Treatment includes primary resection of the tumor and adjuvant therapy or neoadjuvant chemotherapy, resection of the tumor and adjuvant therapy.

Key words: rhabdomyosarcoma; embryonal rhabdomyosarcoma; vaginal bleeding; uterine corpus tumor; tumor of the genitourinary system

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INTRODUCTION

Embryonal rhabdomyosarcoma (RMS) is the most common malignant tumor of the genitourinary system in children. In adolescents it is mainly found in the cervix and uterus, while in infants it is more likely to affect the vagina. The most common symptoms in girls include vaginal bleeding, abdominal pain, urinary and/or stool incontinence and hematuria. Diagnostics, apart from the interview and physical examination, should be based on magnetic resonance imaging (MRI), but the final diagnosis can be made only after the result of the histopathological examination. Treatment includes primary resection of the tumor and adjuvant therapy or neoadjuvant chemotherapy, resection of the tumor and adjuvant therapy.

CASE REPORT

A 13-year-old girl was hospitalized due to vaginal bleeding that continued for 3.5 months in the Department of Surgery and Urology Upper Silesian Child Health Center in Katowice. Then she was transferred to the Department of Pediatric and Adolescent Gynecology in Katowice with the suspicion of a uterine tumor. MRI showed the presence of an enlarged uterus with a contrast-enhancing tumor and lymph nodes enlarged on both sides in the area of the iliac vessels (Fig. 1). Physical examination revealed a distended cervix in association with a hard tumor. The lesion was already investigable near the hymen (Fig. 2). It was decided to take a specimen from tumor for histopathological examination. After the pathomorphological council, the diagnosis of embryonal RMS was obtained.

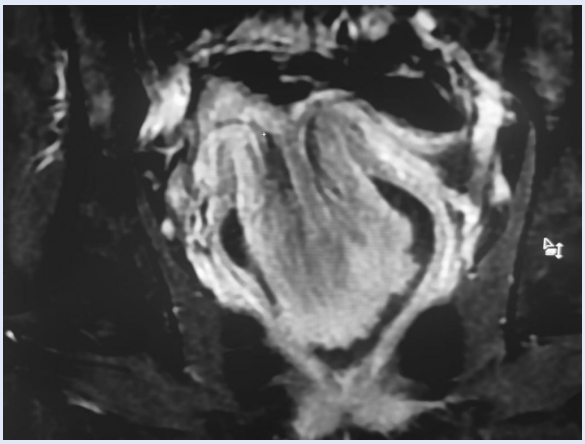

Figure 1. Magnetic resonance image — the bottom of the uterus pulled into the cavity by a tumor

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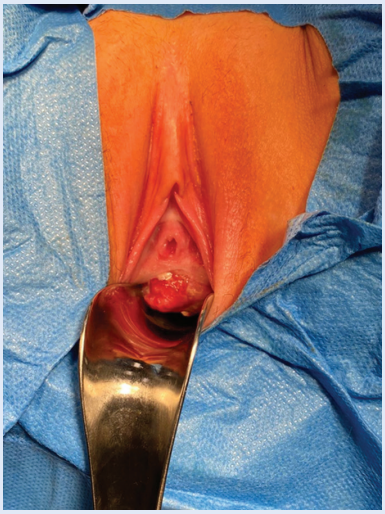

Figure 2. Tumor reaching the hymen

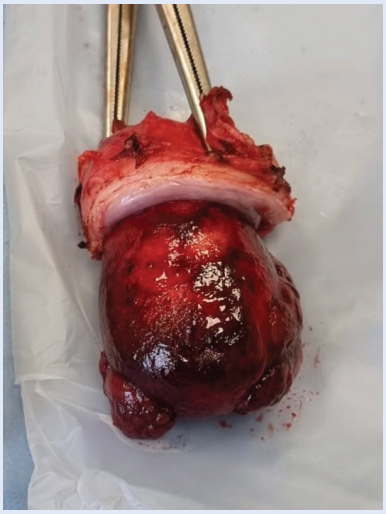

Figure 3. The removed uterus with tumor

Oncologist recommended resection tumor without appendages with pelvic lymphadectomy and adjuvant therapy (chemotherapy or chemotherapy + radiotherapy) afterwards. The procedure was performed as intended (Fig. 3). However, during the operation, there was a complication, which was the discontinuity of the ureters. The patient was treated surgically, a Foley catheter and a ureteral catheter double J 4,0 were inserted after urological consultation. After the laparotomy was performed, the diagnosis was established as Embryonal rhabdomyosarcoma — the stage of disease pT1bN0M0, resection R0.

The patient was transferred to the Department of Pediatric Oncology, Hematology and Chemotherapy of Upper Silesian Child Health Center in Katowice for complementary therapy. She was treated according CWS-guidance protocol, Standard Risk Group (ifosfamide, actinomycinum, vincristinum).

CONCLUSIONS

RMS is a rapidly growing, primitive, high-grade mesenchymal neoplasm [1, 2]. The histological type is an independent prognostic factor. Embryonic tumors have a better prognosis than follicular tumors [3]. The location of sarcoma is also crucial. If the genitourinary system is affected, but the bladder and prostate remain intact, prognosis is most favorable. Imaging tests, especially MRI, are very helpful in diagnosing RMS. Initial diagnosis should allow to run stratification.

The possibility to run a tumor resection depends on the age of the patient and the size and location of the nodular lesion The best healing results are obtained when it is possible to perform a primary tumor resection in the R0 range [4]. In most cases, when the patient is diagnosed the operative procedure is a biopsy [5]. It is recommended to reoperate, if it is possible to remove the microscopic tumor remnants. An important element of treatment is chemotherapy and radiotherapy.

After considering all clinical information about the patient, we can expect a 3-year event free survival (EFS) at the level of probability 70–80% [1].

Conflict of interest

All authors declare no conflict of interest.

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