Rhabdomyosarcoma of The Uterine Cervix: Case Report and Literature Review

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ABSTRACT

Introduction: Embryonal rhabdomyosarcoma (RMS) of the uterine cervix is rare mesenchymal tumor that occurs in less than 1% of all cervical cancers. These malignant tumors occur in adolescents and young adults. It starts from embryonic muscle tissue or pluripotent mesenchymal cells. This case reports an unusual case of embryonal rhabdomyosarcoma in a 33-year-old woman.

Case Presentation: A 33-year-old woman came to the Obstetrics and Gynecology Department. She complained of exophytic cervical masses and vaginal bleeding. There were no remarkable findings about medical, family, and social history. Histopathology confirms embryonal rhabdomyosarcoma (sarcoma botryoides) of the uterine cervix. A combination of radical hysterectomy and adjuvant chemotherapy was chosen as the therapeutic option in this case.

Conclusion: Although the prevalence of cervical rhabdomyosarcoma in adults is very low, a multimodality approach is necessary for favorable prognosis.

INTRODUCTION

Rhabdomyosarcoma is the most common soft tissue sarcoma in children and adolescents. It starts from embryonic muscle tissue or pluripotent mesenchymal cells [1]. This malignant skeletal muscle tumor histologically resembles normal fetal skeletal muscle before innervation. There are four types based on histology, namely embryonal, alveolar, botryoidal, and pleomorphic [2].

Epidemiologically, the incidence of RMS is 4.3 cases in one million children, and about 350 new RMS cases are diagnosed yearly [3]. Previous studies diagnosed fifty percent of RMS patients before the age of five. RMS sites are mainly the head and neck region (25%), genitourinary tract (22%), and extremities (18%), where other observed sites include the thoracic wall, perianal/anal, abdominal, retroperitoneal, and biliary tract (approximately 60% of all newly diagnosed rhabdomyosarcomas are embryonal type and usually occur in younger children) [4].

Around 40% of all rhabdomyosarcomas occur in the neck and head regions. They may occur in the extremities and genitourinary tract. They occur in three primary head and neck sites, orbit, parameningeal sites (nasopharynx, nasal fossa, paranasal sinuses, infra-temporal fossa, pterygoid fossa, middle ear, and mastoid), and non-parameningeal sites (scalp, face, parotid, oral cavity, oropharynx, larynx, and neck) [5]. Nasal or paranasal rhabdomyosarcoma may present with a gradual onset of nasal obstruction and bloody nasal discharge. Tumors in the ear may present with bloody discharge and persistent otalgia, despite treatment. Polypoid masses may be seen in the ear canal or nasal cavity [6].
Although adults with rhabdomyosarcoma had generally worse survival than children with similar tumors, cervical botryoid rhabdomyosarcoma, usually occurring in the second decade has a higher level of survival and better prognosis than vaginal lesions with a peak of incidence in infants [7]. The survival rate of vaginal and cervical lesions were 96% and 60%, respectively. Rhabdomyosarcoma was rare in adults, accounting for 3% of all soft-tissue sarcomas. Embryonal rhabdomyosarcoma of the uterine cervix in adults was rare [8]. This case reports an unusual case of embryonal rhabdomyosarcoma in a 33-year-old woman.

CASE PRESENTATION

A young adult woman aged 33 years old complained of vaginal bleeding since six months before being admitted to the hospital. Patients complained of vaginal bleeding with slimy smelly discharge. There was no history of abnormal bleeding before. The patient had no complaints of fever and pain. There were no remarkable findings about medical, family, and social history. She had her first menstruation at 13 years old with a regular cycle. She was married once time and had 2 children. There was no history of family planning and no history of cancer in her family. On the gynecological examination, we obtained a cervical mass of 8x8x8 cm, involving the upper 1/3 of the vagina without the parametrium. In January 2018, she was carrying the results of a cervix biopsy with sarcoma results suspicious for embryonal rhabdomyosarcoma. The patient was diagnosed with cervical carcinoma IIA and underwent a radical hysterectomy. Post-operative mass biopsy and histopathological examination revealed an embryonal rhabdomyosarcoma (sarcoma botryoides) throughout the cervix.

Uterus size 10x5x4 cm attached one adnexa measuring 3.5x2x1.5 cm. The cervix was 6 cm polypoid, resembling a cluster of grapes, dense but soft. Some other tissues were friable, like fish meat. The endometrium showed no abnormalities. The myometrium was solid, and there was no visible tumor. Histopathological examination (03/02/2018) of the specimen showed in Fig.1. There were no visible tumor cells at the incision edge and vaginal cuff. We obtained no necrosis or bleeding, endometrium glands secretion phase, myometrium solid fibroids, and no tumor cells. The ovaries were present corpus luteum, tubal within normal limits. There were no visible tumor cells at the right and left pelvic lymph nodes.

Postoperatively, the patient continued treatment with Carboplatin Paclitaxel zzzzadjuvant chemotherapy six times until June 2018 and was declared with No Evidence of Disease. The patient continues routine control to the present. At post fourth chemotherapy on 21st June 2018, it was declared a non-evident disease. The patient continues to control the hospital until December 2019 every 3 months. After that, the patient returned for control every 4 months and it was stated that there was no remaining tumor.

DISCUSSION

Rhabdomyosarcoma is a primary soft tissue cancer that starts with the development of immature mesenchymal cells and can become any part of the body except bone. Many cases involve young patients. For example, the study by Amalia, D. showed that patients

Fig. 1. Histopathology of cervical lesion at 10x magnification; (B) Hyperchromatic nucleus with medium-sized, oval, and spindle cells (400x magnification). Histopathological examination revealed a thin polypoid epithelium tumor but intact; the stroma showed variable cellularity, and there was a cellular part whole by a spread of medium-sized cells with round, oval spindle fat, solid chromatin nucleus, and thin cytoplasm.
with RMS were 1-9 years old [3]. Other studies also showed an increased prevalence of embryonal subtype rhabdomyosarcoma in younger patients compared to the alveolar subtype [9]. However, case data in adults are sparse, and rhabdomyosarcoma of the cervix, in particular, has not been widely reported [10].

The case above describes the rhabdomyosarcoma subtype sarcoma botryoides. Botryoides subtype occurs in 6% of rhabdomyosarcomas and belongs to the embryonal rhabdomyosarcoma variant [11]. However, this variant rarely occurs in the cervical region. Only 0.5% of rhabdomyosarcoma subtype botryoides were found in the cervical region. In addition, the study by Kriseman, M., and Ricciardi, E. only found 10-11 cases over ten years [11,12].

The clinical presentation usually shows signs of vaginal bleeding and masses of various sizes in the vagina [13]. However, other symptoms, such as hematuria or abdominal pain, may occur when the tumor is pushed into the abdomen [14]. Vaginal bleeding is the most common presentation of rhabdomyosarcoma. The botryoides variant mass appears like round clusters of grapes [15]. It is similar to our case. The patient showed a polyloid tumor, edematous like grapes, solid but soft, located in the cervical and vaginal areas with a size of 8x8x8 cm. Computed Tomography (CT-Scan) or Magnetic Resonance Imaging (MRI) methods can help to investigate the diagnosis. The mass is also tested for biopsy and histopathological examination to get a definite diagnosis, which is the gold standard in tumor examination [16].

Histopathological features usually show the presence of a layer called “cambium,” a dense layer of primitive neoplastic cells located beneath the epithelial layer, separate from the myxoid, hypocellular zone [17]. However, several other cases also showed pleomorphic spindle-shaped cells [15,18]. In this case, the histopathological features also showed the same results in spindle-shaped pleomorphic cells with hyperchromatic nuclei indicating an active cell division process.

The staging of rhabdomyosarcoma involves three main processes. It consists of tumor staging (TNM; Tumor, node, metastatic classification of malignant tumors), surgical pathology classification using the IRS (Intergroup Rhabdomyosarcoma Study), and risk grouping (High risk, intermediate, and low) to determine the disease management processes in the future [19].

Treatment of rhabdomyosarcoma involves a variety of multimodality approaches. Systemic chemotherapy should be administered together with local therapy to control the size of the mass. Local therapeutic may consist of surgery, radiotherapy, or both to maximize the effectiveness of treatment. Primary resection can be performed before chemotherapy, especially if the mass endangers the patient or causes fatal organ damage. It can be done if the mass has negative margins, such as a para testicular tumor [20]. The chemotherapy generally uses Vincristine (< 1 y: 0.025/kg 1–3 y: 0.05 mg/kg >3 y: 1.5 mg/m2 (max dose 2 mg)), Actinomycin (< 1 y: 0.025/kg > 1 y: 0.045 mg/kg (max dose 2.5 mg)), and Cyclophosphamide (< 1 y: 36 mg/kg 1–3 y: 73 mg/kg > 3 y: 2.2 g/m2) as the first line of treatment for rhabdomyosarcoma.

Several chemotherapy drugs such as Carboplatin/ Etoposide, Ifosfamide/ Carboplatin/ Etoposide, Vincristine/ Irinotecan, Cyclophosphamide/ Topotecan, and Vinorelbine/ Cyclophosphamide are used for patients with recurrent rhabdomyosarcoma [16]. The previous study recommended a total hysterectomy for patients with masses in the cervix and uterus. But, it is not recommended if there is no parametrical involvement [21,22]. Age, unfavorable tumor site, bone marrow involvement, and three metastatic sites can worsen the patient's prognosis [16]. The patient in the case showed the presence of a mass in the vaginal and cervical areas. Thus, a total hysterectomy was performed with the administration of Carboplatin Paclitaxel 6 times to prevent a recurrence.

**CONCLUSION**

The researcher showed a case where total hysterectomy and the administration of Carboplatin Paclitaxel in an adult patient with cervical rhabdomyosarcoma showed favorable outcomes. Although the prevalence of cervical rhabdomyosarcoma in adults is very low, a multimodality approach is necessary for favorable prognostic.

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**CONFLICT OF INTEREST**

The authors declare there is no conflict of interest.

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