Case Report

BOTRROID Embryonal Rhabdomyosarcoma with Uterine Cervix in a Postmenopausal Woman: An Unusual Case Report

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ABSTRACT

Rhabdomyosarcoma (RMS) is one of the most common soft-tissue sarcomas that engage the embryonal skeletal muscle cells as the female reproductive tract. Embryonal RMS (ERMS) is the most prevalent subtype of RMS in the female genital tract. Botryoid RMS is a rapidly growing rare malignancy and a polypoid variant of ERMS that occurs in childhood and constituting approximately 3% of all RMSs among young children and 1% among adolescents and young adults. A 50 year old menopause woman who had been vaginal discharge and bleeding for about 2 years without dysuria, dyspareunia, or postcuital bleeding was informed consent for presenting. A vaginal examination, pathology examination, sonography, magnetic resonance imaging, immunohistochemistry, surgery and radical hysterectomy, radiation therapy, and two sessions of brachytherapy were performed. After 22 months of follow-up, the patient had no evidence of recurrence or any problem in sexual activity. Oncological surgical treatment based on the carcinoma site and adjuvant chemotherapy is helpful for the treatment of RMS. However, applying the standard treatment guidelines is essential, although it is very scarce and difficult.

KEYWORDS: *Botryoid, embryonal rhabdomyosarcoma, rhabdomyosarcoma, uterine cervix, woman*

INTRODUCTION

Rhabdomyosarcoma (RMS) is one of the most common soft-tissue sarcomas that engage the embryonal skeletal muscle cells as the female reproductive tract.[1,2] Embryonal rhabdomyosarcoma (ERMS) is the most prevalent subtype of RMS in the female genital tract.[1] Botryoid RMS is a rapidly growing rare malignancy and a polypoid variant of ERMS that occurs in childhood and constituting approximately 3% of all RMSs among young children and 1% among adolescents and young adults.[1,2]

The primary site of botryoid RMS is closely related to the age of the patient as it is found in the vagina during infancy and it can happen in the cervix during active reproductive age or it may occur in the corpus uteri after menopause.[3] The vaginal form of botryoid RMS is five times more frequent than the cervical type and have better prognosis than the others.[2] We presented a rare unusual case of ERMS in a 50 year old menopause woman that managed and has a successful treatment after 2 year follow up.

CASE REPORT

A 50-year-old menopause woman who had been vaginal discharge and bleeding for about 2 years without dysuria, dyspareunia, or postcuital bleeding was informed consent for presenting. In the cervical examination, there was a polyp, which was biopsied. The Ethical Committee of Qom University of Medical Sciences approved the ethical consideration of this case by IR.MUQ.REC.1402.083 code at 22 Jul, 2023.

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Pathology result

The pathology result showed an endocervical polyp with ulceration and inflammation and negative for dysplasia or malignancy and polypectomy.

Sonography

The dimensions of the uterus were 62 mm × 25 mm × 32 mm in sonography and the ovaries were normal. The heteroechoic mass 45 × 25 × 20 contained vessels that filled the cervical canal and extended to the upper part of the vaginal canal. There was no evidence of the extension of this mass to the uterine cavity.

Magnetic resonance imaging

Abdominopelvic magnetic resonance imaging (MRI) reported that there was a 78 mm × 68 mm × 70 mm mass in the cervix. It showed a heterogeneous increased signal on T2-weighted image and heterogenic enhancing cystic, necrotize components. The post GD images extension into the proximal portion of vagina shows well defined T2 hypointense rim with bulge toward the parametrium and suspicion of parametrial invasion. The mass contained well-defined border with the uterus. However, mild widening of the endometrial cavity at the body of the uterus is visible with a maximum anteroposterior diameter of about 10 mm. Both ovaries were atrophic and no adnexal mass was seen. Large well defined heterogeneously enhancing mass at the uterine cervix is related to a carcinosarcoma with mild extension to the proximal endovaginal canal, and suspicion of early evidence of parametrial invasion [Figure 1]. The patient underwent curettage again; the result of endometrial curettage after 6 months was a fragment of endocervical polyps with chondroid metaplasia. Therefore, cervix adenosarcoma with heterologous elements chondroid differential diagnosis was done. The histological grade was low grade [Figure 2].

Immunohistochemistry

The immunohistochemistry (IHC) was done and its results showed [Figure 3] positive desmin, Myo D1, and cytoplasmic staining WT 1 and negative smooth muscle actin (SMA) with a diagnosis of ERMS (BOTRROID type) of the uterine cervix. IHC was positive results in CK, CYCLIN, DESIM, and PAX8 and negative in C-KIT, SMA, GATA-3, CD10, NAPSIN-A, and ER factors. Carcinoma with heterologous component ERMS was our final diagnosis [Figure 4]. The patient underwent four sessions of neoadjuvant chemotherapy with AIM, doxorubicin, ifosfamide, and mesna [Figure 5]. Then, the patient underwent surgery, so radical hysterectomy, salpingo-oophorectomy, lymphadenectomy, and omentum biopsy were done on June 2021. After the surgery, two sessions of brachytherapy with intracavitary technique and 28 sessions of radiotherapy with ISO

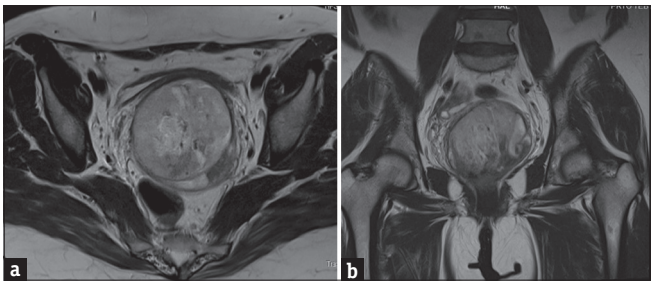

Figure 1: A heterogeneously enhancing mass at the uterine cervix related to known carcinosarcoma with mild extension to the proximal endovaginal canal. (a) Axial image and (b) Lateral image

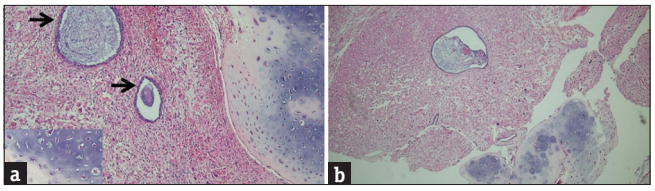

Figure 2: (a) Rhabdomyoblast is infiltrated in the cervical stroma, nonneoplastic endocervical glands are seen in between (arrows), and cartilaginous lobule (heterologous element) with nuclear atypia is seen in the right side. H and E, ×100. Atypical nuclei of hyaline cartilage with high magnification (inset) H and E, × 400, (b) Rhabdomyosarcoma with label of hyaline cartilage (heterologous element) and periglandular cuffing H and E, × 400

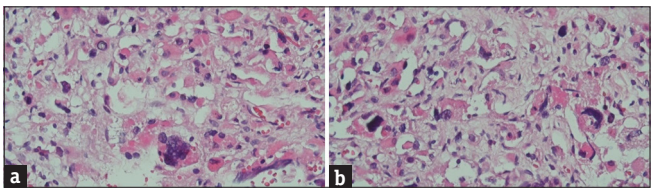

Figure 3: (a) Nuclear pleomorphism with abundant, eosinophilic cytoplasm, and one highly anaplastic multinucleated bizarre tumoral cell H and E, × 400, (b) Anaplastic rhabdomyoblasts were found in several foci. H and E, × 400

gray system and conformal technique with 5040CGY were done. Furthermore, one session of adjuvant chemotherapy was done.

The pathological result after the surgery showed embryonic RMS with heterologous element [Figure 6]. Tumor size was at largest dimension of 5 cm with an additional dimension of 3.5 cm × 3 cm. The tumor site was endocervix with extension to exocervix. Cervical stromal involvement was more than 50% of cervical thickness. No lymph node vascular invasion was seen. Endometrium showed cystic atrophy and myometrium had no specific pathological change. Omentum was mature adipose tissue with free from involvement that observed in the left and right obturator lymph node dissection.

MRI was conducted on March 2022 for comparison with previous MRI with and without contrast and showed salpingohysterectomy and bilateral oophorectomy with no evidence of residual tumor at the surgical site and a

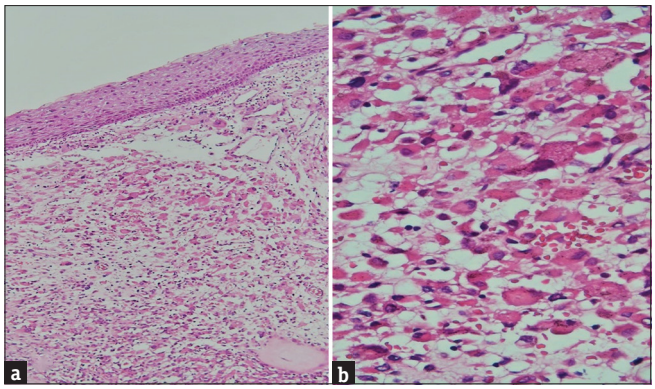

Figure 4: (a) Rhabdomyosarcoma with cellular condensation underlying squamous (cambium layer) H and E, × 100, (b) Primitive round cells with abundant eosinophilic cytoplasm. The rhabdomyoblastic nature of the tumor cells is determined by the small rounded to spindle-shaped tumor cells with eccentric nuclei and eosinophilic tails of eosinophilic cytoplasm (magnification of inset) H and E, × 400

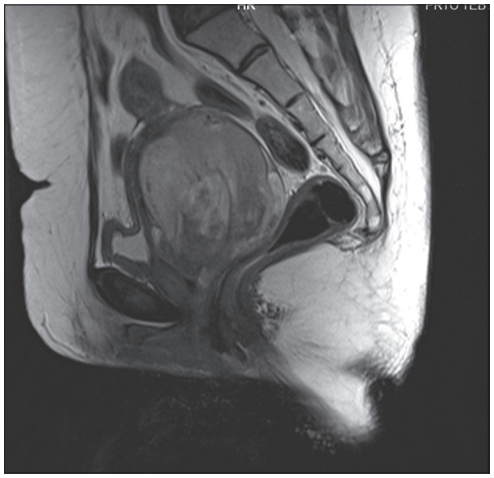

Figure 5: Decreasing size of uterine cervix sarcoma with downward extension representing respond to treatment

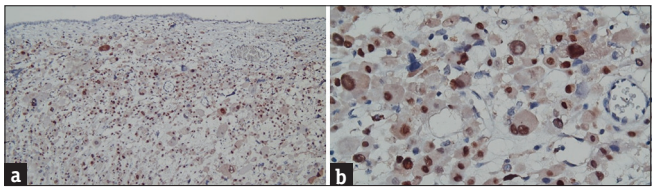

Figure 6: Immunohistochemistry staining for myogenin labeled rhabdomyoblastic cell nuclei. (a) H and E, × 100 high magnification of tumor cells nuclei stained with myogenin, (b) H and E, × 400

small amount of free fluid in the pelvic. No significant pelvic lymph node was seen. A small amount of free fluid was noted in the pelvic cavity. Uniform wall thickening was notable in the rectum, which is probably due to the past radiotherapy changes.

Follow-up and final outcome

The patient's follow-up with physical examination and imaging review during 2 years showed no evidence of progression or recurrence.

Discussion

RMS is occurred in different sites including the head and neck, lymph nodes, extremities, trunk, and the genitourinary tract.[4] From all 20% of RMS in the genitourinary tract of children, the cervix is a rare site of the disease even in childhood and adulthood that engaged the wall of the bladder or vagina.[4,5] Embryonal, alveolar, sclerosing, and pleomorphic types are four different types of RMS tumor, histologically. The embryonal is the most common histology subtype that has three subtypes including botryoid, spindle cell, and not otherwise specified.[5,6]

Botryoid ERMS is a common sarcoma in childhood and adulthood but, the occurrence of this disease in a postmenopausal woman is very rare. Based on the surveys, nearly 90% of all RMS cases are observed in people lower than 25 years of age.[5] In addition, more than 60% of RMSs in this age category occurred in children who have <10 years of age. However, RMS is responsible for 3%–4% of all childhood cancers, but the incidence rate of RMS in adults is lower than 1%.[5,7] In another study, from 115 patients with cervical ERMS, 8.7% were women older than 40 years.[7]

The etiological factors of RMS are not well identified, but low socioeconomic status, recreational drugs consumption during pregnancy and utero radiation exposure are probable factors.[8] According to the recent genetic studies, the frequent chromosomal translocation t (2;13) (q35;q14), involving PAX3 and FKHR genes, may have been related to RMS etiology since these changes are consistently detected in alveolar RMSs.[9] Moreover, it seems that RMS creates from the malignant conversion of immature basic mesenchymal cells and this carcinoma develops in anatomical sites without skeletal muscle.[10]

Treatment of RMS patients is very critical due to the importance of maintaining the reproducibility, fertility conserving, and uterine for child bring in the future. A same treated case without any evidence of recurrence after 6 months was reported by Hermoza *et al*. The case was a 19 year old female with botryoid RMS of the cervix that was treated by cervical polypectomy and adjuvant chemotherapy with vincristine 1.5 mg/m^2/day and actinomycin D 0.045 mg/kg/day for 45 weeks.[1] Moreover, another successfully treated case with radical hysterectomy followed by adjuvant chemotherapy who was a 47-year-old woman is reported by Baiocchi *et al*.[7]

Conclusion

Oncological surgical treatment based on the carcinoma site and adjuvant chemotherapy is helpful for the treatment of RMS. However, by consideration of the

rarity and difficult diagnosis of the disease due to its different histologic subtypes, applying the standard treatment guidelines is essential and therefore, it should be advising by a high expert oncogynecologist.

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Conflicts of interest

There are no conflicts of interest.

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