Case Report-2

Rhabdomyosarcoma of Uterine Cervix in a 44 year female: A rare presentation

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ABSTRACT

Rhabdomyosarcoma (RMS) is a common childhood malignant tumour of striated muscle origin. The common sites are head and neck and genitourinary region. Primary embryonal rhabdomyosarcoma of female genital tract in an adult is rare. A 44 year old premenopausal female presented with complaints of bleeding per vagina and irregular menstruation. On examination a soft bluish fragile irregular mass, arising from the uterus and reaching up to introitus was seen. Biopsy was suggestive of uterine sarcoma, further differentiation was not possible. She underwent total abdominal hysterectomy with bilateral oophorectomy, bilateral lymph node dissection and omental sampling. Final histopathology showed embryonal rhabdomyosarcoma, spindle cell type, arising from the junction of lower uterine segment and endocervix. Immunohistochemistry was positive for vimentin, desmin & myoglobin. She was put on combination chemotherapy.

INTRODUCTION

Rhabdomyosarcoma (RMS) is a soft tissue neoplasm arising from primitive embryonal mesenchyma. Embryonal rhabdomyosarcoma is most common subtype. It is generally regarded as a neoplasm occurring during childhood, although there have been a few reports of this neoplasm in adults[2]. Anatomic presentation and the response of embryonal rhabdomyosarcoma to therapy are different from that of childhood embryonal rhabdomyosarcoma.

CASE : A 44 year premenopausal female presented with complaints of bleeding per vagina and irregular menstruation since 2 months. She denied any major illness in the past. Her menstrual cycles have been normal. She was P4 L4 A0. She was referred to this hospital with a suspicion of a cervical tumour. On per vaginal examination 7-8 cm, soft bluish fragile irregular mass, arising from the uterus and reaching up to introitus was seen. Her routine blood investigations and chest x-ray were normal. Ultrasonography of the pelvis showed anteverted uterus with endometrial thickness of 18 mm without any evidence of mass in either adnexa. bHCG was normal. Biopsy from mass was taken and histopathology showed spindle cells with myxoid stroma & vascular proliferation suggestive of uterine sarcoma. She underwent total abdominal hysterectomy with bilateral oophorectomy with bilateral lymph node dissection with omental sampling. Grossly –polypoidal mass measuring 7×4×2cms in size arising from the junction of lower uterine segment and endocervix. Microscopically tumour showed ulceration of cervical mucosa with spindle cell proliferation. It showed zones of increased cellularity (cambium layer) around the entrapped endocervical glands. Immature cartilage was also present focally. Histopathological picture favoured embryonal rhabdomyosarcoma. Lower

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uterine segment was also infiltrated by tumour (less than 1/3 thickness). Ovaries, posterior vagina and fallopian tubes were grossly and histologically unremarkable. Pelvic lymph nodes and omentum were free of tumour (Stage I-p$T_2N_0M_0$). Immunohistochemistry was positive for vimentin, desmin, actin and myoglobin. She was advised adjuvant chemotherapy.

DISCUSSION

Embryonal rhabdomyosarcoma of the female lower genital tract occurs during childhood, but it has been reported infrequently in adults. It has been considered that a primitive mesenchymal cell is the stem cell of the entire range of soft tissue neoplasms, explaining the appearance of rhabdomyosarcoma in sites where rhabdomyosarcoma are normally not found[1]. It must be separated from adenosarcoma and pseudosarcoma botryoides.

Adenosarcoma may be found in young woman and typically displays condensation of stromal cells beneath the surface epithelium and around glands that may resemble the cambium layer of sarcoma botryoides. Adenosarcoma, however, does not form grape like clusters because stroma is more fibrous and lacks the edematous aspect of sarcoma botryoides. Adenosarcoma also displays a prominent leaf-like pattern characterized by numerous epithelial-lined cysts and cleft-like invagination of the surface resembling cystosarcoma of breast. In contrast, if glands are present within a sarcoma botryoides, they are focal and result of entrapment of surface epithelium.

Pseudosarcoma botryoides (edematous mesodermal cervical polyp) resemble sarcoma botryoides but differ in that polyp usually occur in adult woman as solitary small soft fleshy protuberances. They seldom exceed 1.5 cm in diameter. A cambium layer as rhabdomyosarcoma is not seen and there is more uniformity of sarcoma within mesodermal polyps.[3, 4]

Bernal et al[5] reported embryonal rhabdomyosarcoma in a 19 year old female with cervical polyp. Brand et al[6] reviewed twenty-one cases of sarcoma botryoides of the uterine cervix, including four previously unreported cases. The age of the patients ranged from 5 months to 48 years, with a peak incidence in the group aged 14 to 18 years.

During the past two decades ,the outlook for patients, particularly adults has improved because of adjuvant chemotherapy[7]. Kaserer et al[8] reported a case of cervical embryonal rhabdomyosarcoma in IRS stage Ia in a 32-year-old female. After surgical resection and chemotherapy patient was free for 7 months at the time of reporting. Miyamoto, et al[9] also reported embryonal rhabdomyosarcoma in uterine cervix that was an incidental finding in 46 year old women. According to Miyamoto this was the only fourth case reported in details of sarcoma botryoides of the uterine cervix in a patient over the age of 40 years.

This case reminds us that rhabdomyosarcoma can occur in older female[10] and also at uncommon site. According to Intergroup Rhabdomyosarcoma study this patient falls into group IB with expected 4 year progression free survival >85%. However there are no survival data available for RMS of this site and age group.

CONCLUSION

Awareness of this uncommon lesion in this age and site and its clinical implications is important to avoid misdiagnosis.

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