

Uterine Cervix Rhabdomyosarcoma : A Case Report

Sinha K¹, Adhikari H¹, Baidya P²

ABSTRACT

Rhabdomyosarcoma is a malignant tumor that arises from embryonal skeletal muscle cells. It is responsible for 3% of cancer cases among children aged from 0 to 14 and 1% among adolescent and young adult aged from 15 to 19. Embryonal Rhabdomyosarcoma is the most prevalent subtype in the female genital tract and Botryoid sarcomas being a polypoid variant. In young patients, the majority of genital tract rhabdomyosarcomas occur in vagina; however, the most common site being cervix followed by uterine corpus, in adults. We hereby report a case of 24 years lady P2L2 diagnosed as Botryoid Embryonal Rhabdomyosarcoma with evidence of anaplasia without extrauterine metastasis. Embryonal cervical rhabdomyosarcoma is a rare malignant tumor and its typical manifestations are vaginal bleeding and a pelvic mass. However, attention should also be paid to any abnormal vaginal discharge in young unmarried women. Diagnosis of pure rhabdomyosarcoma in uterus involves perfect sampling and histopathological evaluation.

Keywords: Adolescence, Cervix, Embryonal Rhabdomyosarcoma

Authors:

1. Dr. Kavita Sinha
2. Dr. Home Nath Adhikari
3. Dr. Pooja Baidya

¹Department of Obstetrics and Gynaecology, Nepalgunj Medical College and Teaching Hospital, Kohalpur, Banke, Nepal

²Department of Pathology, Nepalgunj Medical College and Teaching Hospital, Kohalpur, Banke, Nepal

Address for Correspondence:

Dr Kavita Sinha
Associate professor
Department of Obstetrics & Gynecology
Nepalgunj Medical College Teaching Hospital
Kohalpur, Banke, Nepal
Email: ksinhapandey@gmail.com

INTRODUCTION

Rhabdomyosarcoma is a malignant tumor of skeletal muscle-derived cells that usually occurs in children and adolescents. Rhabdomyosarcomas are currently divided into four histological subtypes: (i) embryonic rhabdomyosarcoma; (ii) alveolar rhabdomyosarcoma; (iii) spindle cell or sclerosing rhabdomyosarcoma; and (iv) pleomorphic rhabdomyosarcoma.¹ Of these subtypes, embryonal rhabdomyosarcoma has been one of rarest forms.² In addition, 30% of rhabdomyosarcomas originate in the genitourinary tract, mainly in vagina; less than 0.5% in cervix.³ The embryonal rhabdomyosarcoma is further sub divided into sarcoma botryoides and anaplastic variant. These are highly malignant tumors and account for 4-6% of all malignancies in children and young adults.^{4,5} These are usually seen in second decade of life. Botryoid sarcomas are a polypoid variant constituting 3% of all rhabdomyosarcoma. Management of cervical rhabdomyosarcoma ranges from radical surgery to a more fertility sparing conservative approach, along with chemotherapy and radiotherapy. We report here a case of Embryonal Rhabdomyosarcoma of botryoids variety.

Case report

We report here a case of 24 years lady P2L2, referred to our center for suspected Malignant Cervical mass. On elaborating the history, she had developed something coming out per vagina three months back which was small and reducible initially but later the mass increased in size and became irreducible. She had difficulty in micturition and defecation as well. Our provisional diagnosis was Malignant fibroid polyp / Uterine Inversion.

Incisional Biopsy was done and send for histopathological examination which revealed infarcted inflammatory lesion likely polyp and MRI pelvis was suggestive of uterine prolapse, heterogenous fungating mass protruding from the introitus possibly originating from the posterior endocervical wall likely prolapsed polyp. The excision of the cervical mass was planned but during surgery on blunt dissection the whole uterus delivered out probably due to lax pelvic floor muscle, hence vaginal hysterectomy was done. Postoperative period was uneventful.

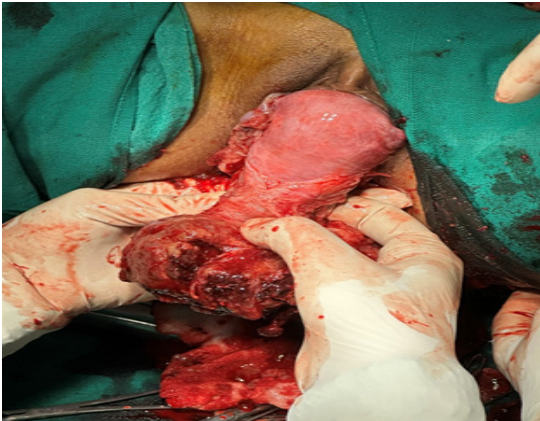


Figure 1

Above pictures describes preoperative and intraoperative appearance of rhabdomyosarcoma of uterus of 24 years lady.

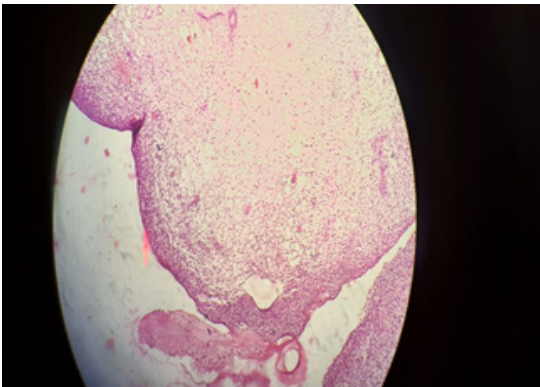


Figure 2: Polypoidal area showing distinct subepithelial condensed layer of tumor cells (cambium layer) separated by loose myxoid stroma

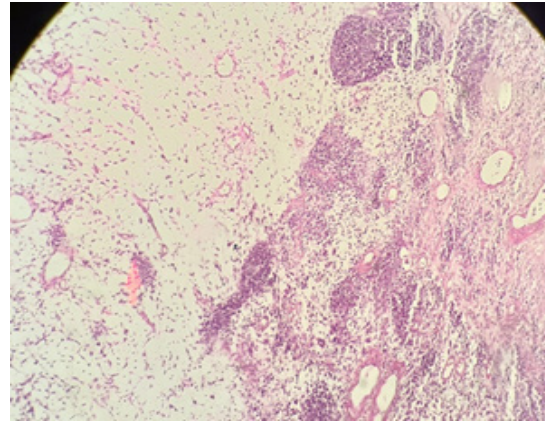


Figure 3: Showing medium power view showing densely packed hypercellular areas composed of small undifferentiated cells adjacent to hypocellular myxoid area on the left

The histopathology examination revealed Botryoid Embryonal Rhabdomyosarcoma with evidence of anaplasia without extrauterine metastasis as shown in figure 2 and 3. Patient was referred to cancer hospital for the opinion about adjuvant treatment.

DISCUSSION

Sarcoma botryoides accounts for 10% of the RMS cases. These embryonal tumors arise from the vagina, cervix and bladder. The outcome is favorable compared to the other subtypes-alveolar and undifferentiated.⁶ Classical embryonal RMS most commonly present with polypoid (exophytic) growth pattern associated with more favorable prognosis (92% survival at 10 years) than diffuse intramural (endophytic) growth pattern (68% survival at 10 years). Botryoid subtype appears on the mucosal surface of hollow viscera such as vagina, bladder and cervix which predominates in children and young adults. Most patients experience vaginal bleeding or a mass sensation at the introitus.⁷ The most important prognostic factors for candidacy to fertility sparing surgery are well defined (polypoid) presentations, embryonal histology, superficial tumors, whilst deeply invasive disease, alveolar/pleomorphic histology increase the risk of treatment failure and tumor recurrence.⁸ Also, the extent of the disease after primary surgery is another important prognostic factor. In addition, rhabdomyosarcoma of the female genital tract, especially the uterine corpus and rarely the uterine cervix, may be associated with a pathogenic DICER1 gene variation.⁹ This is important because the detection of this mutation in a patient or relatives can provide the opportunity for surveillance of related conditions that might improve long-term outcomes and survival.

CONCLUSION

Embryonal rhabdomyosarcomas of the uterus are extremely rare malignant tumors. Its occurrence in women over 20 years of age is rare, and studies on treatments and outcomes are limited. However, attention should also be paid to any

abnormal vaginal discharge in young unmarried women.

REFERENCES

1. Leiner J, Le Loarer F. The current landscape of rhabdomyosarcomas: an update. *Virchows Arch* 2020; 476: 97–108.
2. Suneja A., Malik R., Guleria K., Vaid N. B., Varma D. D., Mishra K. Embryonal rhabdomyosarcoma causing uterine inversion in an adolescent: a rare tumor with rare association. *Indian Journal of Gynecologic Oncology*. 2020;18(4):1–3. doi: 10.1007/s40944-020-00474-9.
3. Ricciardi E, Plett H, Sangiorgio V, et al. Adult primary cervical rhabdomyosarcomas: A Multicentric cross-national case series. *Int J Gynecol Cancer* 2020; 30: 21–28.
4. S Jayi et al. Embryonal rhabdomyosarcoma of the cervix presenting as a cervical polyp in a 16-year-old adolescent: a case report. *Journal of Medical Case Reports* 2014; July 1:8:241.
5. Kriseman ML, Wang W-L, Sullinger J, Schmeler KM, Ramirez PT, Herzog CE, Frumovitz M. Rhabdomyosarcoma of the cervix in adult women and younger patients. *Gynecol Oncol*. 2012; 126:351–356.
6. F.E. Buruiana, B. Gupta, and K. Singh. Rhabdomyosarcoma of the cervix in teenagers – Is fertility preservation a feasible option?. *Gynecol Oncol Rep*. 2020 Nov 21; 34: 100677.
7. Alejandro Delfos Hermoza a, Graziela de Macêdo Matsushita b, Marcelo Henrique dos Santos c, Ronaldo Luis Schmidt c, Ricardo dos Reis c, Carlos Eduardo Mattos da Cunha Andrade c. Botryoid embryonal rhabdomyosarcoma of the cervix: A case report. *International Journal of Surgery Case Reports*. 2023; 102:107858.
8. Elsebaie M.A., Elsayed Z. Is Fertility-preservation Safe for Adult Non-Metastatic Gynecologic Rhabdomyosarcoma Patients? Systematic Review and Pooled Survival Analysis of 137 Patients. *Arch. Gynecol. Obstet*. 2018; 297:559–72.
9. Kebudi R, Dural O, Bay SB, et al. Childhood Rhabdomyosarcoma of the Female Genital Tract: Association with Pathogenic DICER1 Variation, Clinicopathological Features, and Outcomes. *Journal of Pediatric and Adolescent Gynaecology*. 2021;34(4): 449-453.