Cervical Rhabdomyosarcoma; A Rare Entity

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Introduction

- Rhabdomyosarcoma (RMS), a malignant neoplasm arises from skeletal muscle progenitors is frequently seen in pediatric adolescent population.
- In adults, RMS are rare, making < 5% of all soft tissue sarcomas and <1% of all malignancies while RMS of the female genital tract occurs in 3.5% of all cases.
- Cervical RMS mostly affects women in their 2nd and 3rd decade of life and of all cervical cancers, RMS incidence is reported to be about 0.4 to 1% .
- WHO, histologic subtypes of RMS with incidence are i) embryonic (59%) with subclass:- classic(49%), botryoid (6%) and spindle cell (3%) ii) alveolar (21%) iii) anaplastic (11%) iv) NOS (8%).
- Given the rarity of clinical situation, there are limited data, mostly retrospective single institute studies, available on RMS in adults

Figures

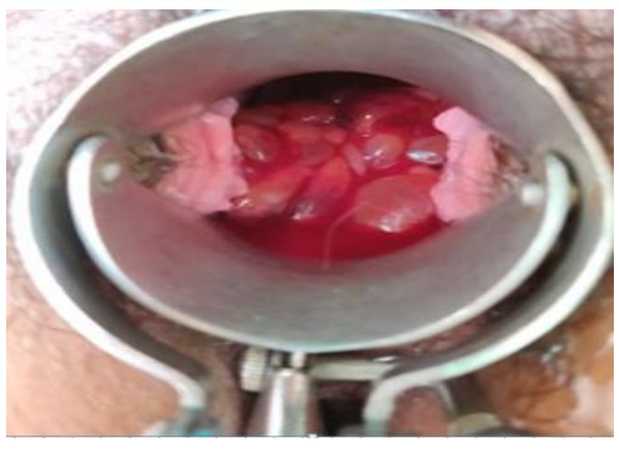

Fig 1A

Fig 1B

Figure 1. Showing vaginal speculum examination: Multiple polypoidal growth from cervix which bleeds on touch about 6x4cm in Fig 1A and after chemotherapy 3x3 cm size in Fig. 1B

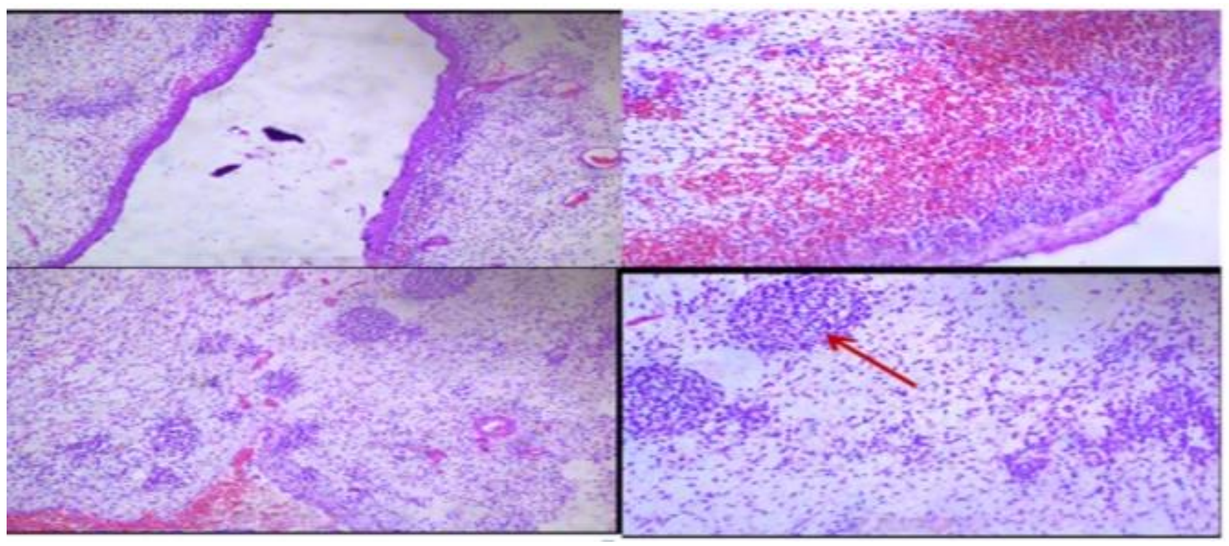

Figure 2. Showing the section of polypoid tissue lined partly by endocervical columnar epithelium and partly by stratified squamous epithelium. Subepithelial dense aggregates of rhabdomyoblasts (the so-called "cambrium" layer) are characteristic. Occasional Mitosis. IHC showed positive for Myogenin and Desmin.

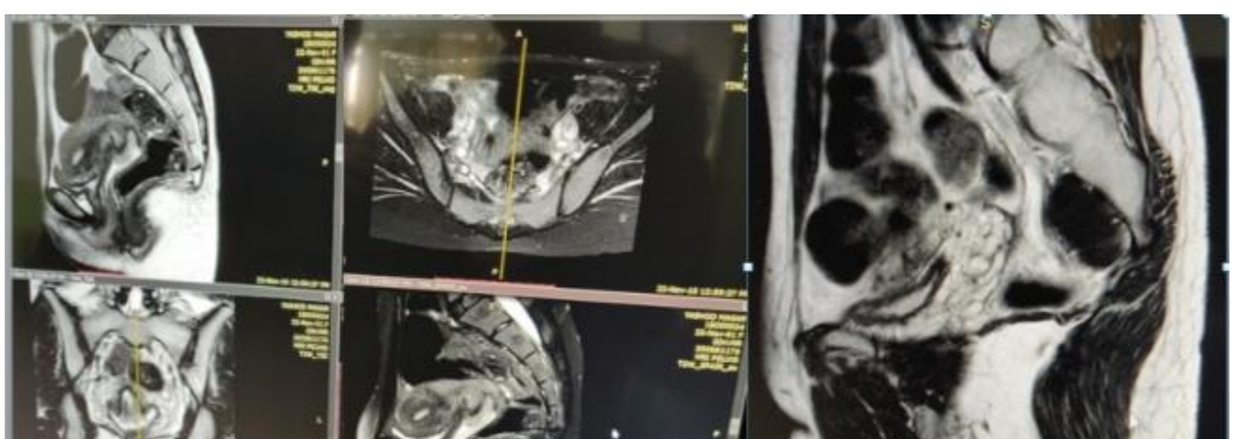

Figure 3. MRI pelvis shows Lobulated cystic lesion displaying iso to low signal in T1 and high signal in T2 and fat suppressed images noted in cervical canal and upper part of vaginal canal measuring 6.5x 5.3 cm in size. Cervical stroma is normal. Rest of the uterus shows normal morphology and signal intensity.

Case Summary

- Mrs. Y. M, 27yrs, P1L1 was referred to our Gynecologic-oncology OPD on 13th Jan 2019 with a diagnosis of Cervical Rhabdomyosarcoma: Botryoid variant on LEEP biopsy and confirmed by IHC.
- She had undergone cervical polypectomy four times in last one year which was previously reported as cervical polyp without atypia.
- **Our plan of treatment after MDT Discussion was Chemotherapy** with VAI (Vincristine, Actinomycin-D and Ifosphamide) and **re-assess after 3 cycles** for Treatment response and further management with radical Surgery with pelvic lymph node dissection followed by adjuvant Chemotherapy. Consider for RT if nodes and/or LVSI positive.
- After 1st cycle chemotherapy there was 50% regression in tumor size

Discussion

- Genital tract RMS primarily seen in prepubertal girls and adolescents, has excellent survival rates; however, outcomes for adults remain poor.
- Treatment of RMS in adults is usually multimodality (surgery, radiotherapy and chemotherapy).
- Studies done in MD Anderson Cancer Center and Dana Faber Cancer institute have reported high overall response rate 75% and 82% respectively and better survival with disease responding to chemotherapy (10yrs and 5yrs survival 72% and 57% respectively) with standard VAC (C-Cyclopdosphamide) regimen.
- Better 5yrs survival of 63% have been shown in those with complete resection of disease in contrast to those with incomplete resection.
- For localized disease, chemotherapy combined with locoregional modality (surgery/radiotherapy) cure rates of ~ 90% can be achieved.
- Neoadjuvant chemotherapy can greatly reduce tumor burden and permit an organ preserving surgery.
- Genetic link between cervical RMS and other primary tumors, most notably Sertoli–Leydig tumor have been reported as some studies have noted DICER1 germline mutation.

Conclusion

Cervical RMS in adults is rare disease with poor outcome. So to achieve better cure rate, treatment is usually multimodality.

References

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