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Embryonal Rhabdomyosarcoma Yolk Sac Tumour of the Uterine Cervix: A Case Report ofClear Cell Adenocarcinoma and a Discussion of Its Line of Treatment

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ABSTRACT:[1]Embryonal rhabdomyosarcoma of the uterine cervix is an uncommon presentation of the most common soft-tissue sarcoma in the first decades of life.Rhabdomyosarcoma is the most common soft-tissue sarcoma of childhood, accounting for 50% of those cases diagnosed at or before 20 years of age.One of the least common sites for rhabdomyosarcoma in the genitourinary tract is the uterine cervix.[2]Malignant germ cell tumours are rare tumours of childhood accounting for less than 3% of paediatric malignancies, with endodermal sinus tumour (EST) being the most common histological subtype. The vagina is an extremely rare site for primary germ cell tumours.Clinically, it is commonly diagnosed as botryoide rhabdomyosarcoma. We at Gujarat cancer and research institute report a case of rhabdomyosarcoma, primary yolk sac tumour which has histopathological characteristic of clear cell adenocarcinoma.

KEYWORDS: Rhabdomyosarcoma,Clear Cell Adenocarcinoma,Endometrial sinus tumour, Botryoiderhabdomyosarcoma

I. INTRODUCTION

[3]Cervical carcinoma is the most common malignancy in Indian women and fifth most common cause of cancer related death. Squamous cell carcinomas are the largest subset, whereas adenocarcinomas constitute only 15% of cervical carcinomas.[2] Clear cell carcinoma of cervix is a very rare malignancy accounting only 4% of all adenocarcinoma of cervix. Intrauterine exposure to diethylstilbestrol (DES) is supposed to be causative factor for clear cell adenocarcinoma in childhood and young-age patients.Clear cell carcinoma of cervix commonly presents with abnormal per-vaginal bleeding (menorrhea, menometrorrhea, post-menopausal bleeding) refractory to hormonal therapy. Most of the clear cell carcinomas are superficial with invasion to deeper stroma rather than exophytic mass forming lesions.In children, it also to be differentiated from yolk sac tumour and embryonal rhabdomyosarcoma. Unfavourable prognosis is related with larger size, higher stage, high mitotic rate, positive surgical margin, parametrial involvement and lympho-vascular spread.Neo-adjuvant chemoradiation are indicated for these cases. Metastasis is uncommon but local recurrence may occur.

CASE REORT

A 1 Year old female with no comorbidities with h/o NICU admission for jaundice, presented with per vaginal painless bleeding since 25 days. MRI pelvis showed 36 x 36 x 56 mm sized lesion involving uterus and vagina. There was extension of lesion anteriorly indentation over posterior wall of urinary bladder and superiorly capsular breech with focal loss of fat plane with bowel loops. Uterus was not seen separated from lesion Histopathological(biopsy) examination from material passed out of vagina s/o clear cell adenocarcinoma. Lesion appears hypointense on TLW. Her AFP was 7781 ng/ml,β HCG was 0.1 and LDH was 238.7. Her s2 marker was suggestive of grade IIIB. With diagnosis of Rhabdomyosarcoma of vagina,she was admitted in female medical ward and immediately given inj cefo sulbactam(cephalosporin antibiotic), inj rantac, inj perinorm, injPCM, inj Tranexa, inj botropase (heamocoagulation).

On referring the patient to gynaecology oncology department,the disease s/o high grade malignant tumour clear cell carcinoma.

Neo adjuvant chemotherapy was suggested for 2 cycle in interest of operability. CT Scan thorax was done, and it was in normal limits. Further line of treatment was suggested by paediatric surgeon to insert HICKMAN'S INSERTION with 3rd cycle JEB. HICKMAN'S INSERTION WAS DONE AS PER PLAN.

She was kept on observation.

1st chemotherapy was given (09/01/2018)

DRUG	DOSE	DAYS
CARBOPLATIN	20 MG/ M^2 /DAY	1,2,3,4,5

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ETOPOSIDE	32 MG IN 250 ML DNS IV OVER 2 HOURS	1,2,3,4,5
INJ BLEOMYCIN	4 UNITS SLOW PUSH	2,9,16
INJ EMSET + INJ DEXONA	8MG AND 8 MG RESPECTIVELY IN 100 ML NS OVER 30 MINUTES	1,2,3,4,5

TOLERATTED WELL
values of AFP after chemo was 4.68 AND β HCG WAS 0.1
Kept on observation.

Paediatric opinion was no surgery required. Regular follow up needed.

On 21/07/2018 with parent's consent HICKMAN'S INSERSION WAS DONE IN OT.

2nd chemotherapy was given (24/07/2018)

DRUG	DOSE	DAY
CISPLATIN	6 MG IN HALF PINT OVER 2 HOURS	1,2,3,4,5
IFOSFAMIDE	0.36 GMS IN 250 ML RL OVER2 HOURS	1,2,3,4,5
MENSA	1/3 AMPULE	AT 0,4,8 HOURS OF IFOS
VINBLASTIN	0.04 MG IV SLOW PUSH IN RUNNING IV LINE	1, 2
INJ EMSET + INJ DEXONA	8MG AND 8 MG RESPECTIVELY IN 50 ML NS OVER 30 MINUTES	1,2,3,4,5
ISOLYTE-M + MgSO4	250ML ISOLYTE-M +1 AMPULE OF MgSO4	1,2,3,4,5
INJ MANNITOL 20%	100 ML IV OVER 30 MINUTES	1,2,3,4,5
IV RL	200 ML OVER 2 HOURS	1,2,3,4,5

0n 18/08/2018 there was recurrence of yolk sac tumour posterior to urinary bladder. Patient was admitted and planned for 3rd chemotherapy.

3rd chemotherapy was given (03/09/18)

DRUG	DOSE	DAY
CISPLATIN	7 MG IN HALF PINT OVER 2 HOURS	1,2,3,4,5
IFOSFAMIDE	0.0.4 GMS IN 200 ML RL OVER2 HOURS	1,2,3,4,5
MENSA	1/2 AMPULE	AT 0,4,8 HOURS OF IFOS
VINBLASTIN	0.04 MG IV SLOW PUSH IN RUNNING IV LINE	1, 2
INJ EMSET + INJ DEXONA	10 MG AND 8 MG RESPECTIVELY IN 50 ML NS OVER 30 MINUTES	1,2,3,4,5
ISOLYTE-M + MgSO4	2 PINTS ISOLYTE-M + 2 AMPULES OF MgSO4	1,2,3,4,5
INJ MANNITOL 20%	50 ML IV OVER 30 MINUTES	1,2,3,4,5
IV RL	250 ML OVER 2 HOURS	1,2,3,4,5

After 3rd chemotherapy lesion size was decreased.

On 23/10/2018 again HICKMAN'S CATHETER was removed.

Observation for 3 months.

Again, per vaginal bleeding happened and MRI showed 17 x 15 x 20 mm lesion posterior to urinary bladder. AFP was 622 and β HCG was 0.1. LDH was 218.
CT Scan revealed lung metastasis. Disease became painful as it progresses. Generalized pain due to metastasis.

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4th chemotherapy was planned. (03/01/2019)

DRUG	DOSE	DAY
CISPLATIN	7 MG IN HALF PINT OVER 2 HOURS	1,2,3,4,5
IFOSFAMIDE	0.4 GMS IN 100 ML RL OVER2 HOURS	1,2,3,4,5
MENSA	1/3 AMPULE	AT 0,4,8 HOURS OF IFOS
VINBLASTIN	0.04 MG IV SLOW PUSH IN RUNNING IV LINE	1, 2
INJ EMSET + INJ DEXONA	16 MG AND 8 MG RESPECTIVELY IN 50 ML NS OVER 30 MINUTES	1,2,3,4,5
ISOLYTE-M + MgSO4	250ML ISOLYTE-M +2 AMPULE OF MgSO4	1,2,3,4,5
INJ MANNITOL 20%	50 ML IV OVER 30 MINUTES	1,2,3,4,5
IV RL	250 ML OVER 2 HOURS	1,2,3,4,5

values of AFP after chemo was 67.45 AND β HCG WAS 0.10. LDH was 229
Kept on observation for 3 months.
Regular follow up needed.

Disease recurrence. 5th chemotherapy (06/05/2019)

DRUG	DOSE	DAYS
INJ EMSET + INJ DEXONA	4MG + 4MG IN 100 ML NS OVER 30 MINUTES	1,2,3
INJ AVIL	1CC	1
INJ RANTAC	25 MG IN	1

WAIT TILL 30 MINUTES

DRUG	DOSE	DAY
INJ PACLITAXEL	80 MG IN 250 ML NS GLASS BOTTLE IV OVER 3 HOURS THROUGH CODON FILTERS	ONLY ON DAY 1
ISOLYTE-M +NaHCO3	1 PINT ISO + 1 AMPULE OF NaHCO3	DAY 1
ISOLYTE-M+ MgSO4	1 PINT ISO + 2 AMPULES MgSO4	DAY 2
INJ IFOSFAMIDE	330 GMS IN 250 ML RL OVER 3 HOURS	DAY 1 AND 2
INJ MENSA	1/2 AMPULES	AT 0,4,8 HOURS OF IFOSFAMIDE
IN RL	250 ML OVER 2 HOURS	DAY 1 AND 2
INJ MENSA	1/2 AMPULES	AT 0,4,8 HOURS OF IFOSFAMIDE (DAY 2)
INJ CISPLASTIN	20 MG IN 250 NS OVER 2 HOURS	DAY 2

Patient present with per vaginal bleeding for 10 day and general condition was fair. Noticed pallor.
LIVER METASTASIS(multiple lesion). Both LUNG FIELD shows multiple lesions.
GEMOX 3 cycle was planned but LFU since 5 months.
Patient was sent for radiotherapy for palliative RT opinion and palliative medicine for BLS as her condition seems terminal.

PALLIATIVE RT GIVEN (30 Gy / 10 #) after PAC evaluation.

II. PALLIATIVE MEDICINE

Patient's parents were frustrated and at the edge of crying. Our departmental head did counselling of parents and made them realize the situation(breaking the bad news). Parents were

educated and spiritual and very co-operative. They were counselled about palliative treatment and with their consent line of treatment was designed. Patient was given inj Tranexa immediately as she was bleeding per vaginal and crying endlessly. Parents were taught about role of morphine and how to give it. 10 mg tablet of morphine IR should be crushed and then 10 ml NS was added. The mixer contains 1mg morphine per 1 ml. they were given 10 cc syringe and advised to give 1 ml solution every 4 hours and if needed 2ml solution every 4 hours for pain relief. Syrup loose was given with morphine regimen as it might cause constipation. Tab.Tranexa 500 mg ½ - ½ - ½ till bleeding stops. And then as per requirement.
Diet counselling was also done. After two doses of morphine patient was quite comfortable. Local an

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