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Case Report

Vaginal Rhabdomyosarcoma in a patient with advanced cervical cancer; a case report and review of literature

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ABSTRACT

Rhabdomyosarcoma is very rare in adults accounting for less than 5% of all soft tissue tumours and less than 1% of all malignancies. Vagina is one of the least common sites for occurrence of Rhabdomyosarcoma in the genital tract. We present a case of a 53-year-old woman who is a follow up case of cervical cancer stage IIIB, managed by radiotherapy and chemotherapy. She was doing well till 5 years of her treatment for cervical cancer when she presented with complaints of pain lower abdomen and discharge per vaginum for 10 days. On examination she was found to have an abdominal mass of 18 weeks size and on local examination there was 4X4 cm fixed mass on lower third of vagina arising from left side. MRI abdomen and pelvis was done. Biopsy from the vaginal mass showed features of Rhabdomyosarcoma. Further follow up of the patient was not possible due to lockdown in view of the pandemic. She was last contacted telephonically on 25th March 2020; she said she was waiting for the lockdown to be lifted so that her further management can take place. This is just one patient; there are many more with other medical conditions all over the world who are losing their lives because of not being able to access medical care due to the present pandemic. New growth in the region of local recurrence in a known malignancy cannot necessarily be the recurrence of the primary tumour. It is important to keep our mind open to other differentials apart from the recurrence of primary malignancy, sometimes it can turn out to be a very rare tumour as we encountered in our case.

Keywords: Rhabdomyosarcoma, Soft tissue tumour, Vaginal mass

INTRODUCTION

Rhabdomyosarcoma is a malignant neoplasm arising from progenitors of skeletal muscle. It is the most common soft tissue tumor in children, responsible for approximately 50% of all soft tissue sarcomas and 3% to 4% of all cancers.¹ Most Rhabdomyosarcomas in children occur in the head and neck region; followed by the genitourinary tract.² The tumour is rare in adults, accounting for less than 5% of all soft tissue sarcomas and less than 1% of all malignancies.³ Rhabdomyosarcomas arising in the genitourinary tract are commonly the sarcoma botryoides variant and are typically seen in the first decade of life often before 3 years of age in the case of vaginal rhabdomyosarcoma.^{4,5} One of the least common sites for rhabdomyosarcoma in the genitourinary tract is the uterine

cervix followed by uterine corpus and vagina.⁶ Because of the extreme rarity of vaginal rhabdomyosarcoma, the literature on this disease consists mainly of case reports, and no large case series or randomized trials have been published so far.

We present a rare case report of Rhabdomyosarcoma of vagina diagnosed in a postmenopausal female with cervical cancer Stage IIIB.

CASE REPORT

A 53-year-old female was diagnosed with non-keratinizing squamous cell carcinoma of the cervix 7 years back (July 2013), when she presented with complaints of foul-smelling discharge per vaginum and irregular bleeding per

vaginum (PV). Further investigations by routine blood tests, imaging and biopsy from the mass was done. She was diagnosed with non-keratinizing squamous cell carcinoma of cervix stage IIIB and was started on radiation therapy with concurrent chemotherapy. After completion of her radiotherapy she was on routine follow up and was asymptomatic. In November 2019, she presented to the cancer clinic, 2 months before her routine yearly follow up, with complaints of pain lower abdomen and discharge per vaginum for 10 days. There was no history of bleeding PV, no history of cough, breathlessness, bowel and bladder complaints. On examination, her ECOG performance status was 1, General physical examination was unremarkable and chest and CVS examinations were within normal limits. On per abdominal examination, there was an abdominopelvic mass of around 18 weeks size, firm to hard in consistency, non-tender with restricted side to side mobility, lower limit of the mass was not reached. On per speculum examination, there was a 4×4 cm fixed mass on lower third of vagina arising from left side. Cervix could not be visualized because of post radiation therapy vaginal stenosis. Same abdominopelvic mass was felt on PV examination. On per rectal examination, rectal mucosa and bilateral parametria were free. A clinical impression of recurrence of cervical cancer with uterine and vaginal involvement was made and patient was advised to get a Magnetic Resonance Imaging and a biopsy from the vaginal mass. Contrast enhance MRI abdomen and pelvis was done. On MRI, upper abdomen was unremarkable and uterus was bulky with voluminous collection in the endometrial cavity extending upto the cervical canal (Hematometra) measuring 8.6×10.3×18.9 cm in size, T2W heterointense lesion was seen in the mid segment of endometrial cavity predominantly in the left posterolateral aspect measuring 8×7×7 cm with no serosal extension, Bilateral parametrium were maintained. Biopsy from vaginal mass and endocervical biopsy were taken in the minor Operation Theatre under all aseptic precautions. Biopsy from the vaginal mass showed features of Rhabdomyosarcoma with Tumour cells immune-positive for desmin and myogenin and immune-negative for p40, HMB45, chromogranin and synaptophysin.

Endocervical report was unremarkable with no evidence of dysplasia or malignancy in the sections examined. Ultrasound guided biopsy was taken from the endometrium, as it was not possible vaginally because of extensive stenosis, the report was inconclusive showing coagulative necrosis with few inflammatory cells. With the vaginal mass showing features of Rhabdomyosarcoma, patient was referred back to the cancer clinic for multidisciplinary team management. Patient used to come for follow up to our tertiary care centre from a small village of another state. Soon, COVID-19 was declared a pandemic. Lockdown measures were applied all over the country and further follow up of the patient was not possible. We last contacted her telephonically on 25th March 2020; she was apparently well with the symptom of pain being managed with analgesics, awaiting the lockdown to be lifted so that her management with respect

to the new diagnosis can be commenced. This is just one patient we know of who is suffering not from the pandemic but because of the pandemic, and there are many more with other medical conditions all over the world who are losing their lives because of not being able to access medical care on time.

DISCUSSION

Adult RMS constitutes a very small fraction of soft tissue sarcomas, estimated between 2% and 5%. Partly as a result of the rarity and partly because of difficulties in histologic diagnosis, little is known about the clinic pathologic factors that influence disease recurrence and survival in adult patients with RMS. Urogenital Rhabdomyosarcomas are associated with a more favorable prognosis than tumors in retroperitoneal, limbs, and parameningeal sites.⁴

The accepted treatment modality consists of surgery, chemotherapy and radiotherapy. The aim of surgery should be complete surgical resection with negative margins and preservation of both function and cosmesis wherever feasible.

Several randomized, controlled trials in children have shown that effective adjuvant chemotherapeutic regimens for patients with pediatric RMS have improved the overall survival rates approaching 70%.⁷⁻⁹ The management of adult patients has been extrapolated from the experience with pediatric patients Rhabdomyosarcoma.⁹ Due to paucity of the number of cases in adults, no prospective, randomized trials have been conducted in adults, the available retrospective studies and cases series suggest that the response to treatment is not as good in adults as in pediatric population.

A retrospective review from 1980 to 2010 by Maya et al showed that 11 females presented with cervical rhabdomyosarcoma at a median age 18.4 years, vaginal bleeding was the most common presenting symptom, and a vaginal mass as a co presenting symptom in most of them. Nine patients (82%) received multimodal therapy consisting of surgery with chemotherapy, radiation therapy, or both. 3 patients experienced local recurrence. At a median follow-up of 23 months, 6 patients (55%) were without evidence of disease, 1 (9%) was alive with disease, 1 (9%) had died of disease, and 3 (27%) had died of other causes. Three patients (27%) had other primary malignancies in addition to rhabdomyosarcoma-1 had a sertoli-leydig tumor, 1 had a Sertoli-leydig tumor and a pinealoblastoma, and 1 had thyroid cancer and a parotid adenocarcinoma. The authors concluded that with multimodal therapy, cervical rhabdomyosarcoma appears to be associated with a good prognosis.⁶

The poorer survival seen in adults might be due to the fact that adults have an increased incidence of poor prognostic features as compared to children. A previous series of 84 patients noted that pleomorphic subtype was more

common among adult patients, and was present in 42% of patients greater than age 40.¹⁰

In a retrospective analysis of 25 cases of adult RMS carried at a tertiary institute in India from 2000 to 2009, the 5-year OS rate in this series was 45%.¹¹ In 1970 Hilgers et al. recommended that the best treatment for vaginal RMS would be pelvic exenteration. During the following years, it was demonstrated that preoperative chemotherapy with or without radiotherapy could allow preservation of the bladder and rectum.¹² The treatment modalities have changed over the years with availability of chemotherapeutic agents and use of radiation therapy for management of Rhabdomyosarcomas.

In a study by Hawkins et al, the favorable prognostic characteristics for disease specific survival were age ≤ 20 years, tumors ≤ 5 cm, absence of regional or distant disease, and surgical resection with negative margins.¹⁰

CONCLUSION

A secondary malignancy unrelated to the primary tumour may be a possibility and should be one of the differentials when planning the management. Recurrence of growth is not always recurrence of the primary malignancy, and sometimes it can be as rare as the tumour we came across. Before the introduction of effective adjuvant chemotherapy, the prognosis of Rhabdomyosarcoma was poor. The results of recent trials have shown that the introduction of adjuvant chemotherapy in the management of RMS has made a substantial contribution to the improvement in survival. The mode of management has changed significantly over the past 20 years. In context of our present patient, we do not really know the effect of therapy on her survival, being a secondary malignancy, as the patient and her family await the lifting up of Lockdown, we expect to see some improvement in survival with treatment.

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