Role of Paediatric Palliative Care in a One - Year Old Patient with Yolk Sac Tumour of Vagina and Uterus: A Case Report

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Summary

Yolk Sac Tumour (YST) is a rare and highly malignant germ cell tumour of childhood. Diagnosis is based on histopathological, immune-histochemical studies and highly elevated alfa fetoprotein level. Pediatric oncology population in India does not receive palliative care and only few who do are in late stage disease. Early integration of palliative care improves the quality of life of patients and caregivers throughout the course of illness. We describe role of palliative care in a child with rare tumor

Keywords: Pediatric Palliative Care, Yolk Sac Tumour

Introduction

Malignant Germ Cell Tumour (MGCT) is uncommon tumour of childhood and accounts around 3% of paediatrics malignancies, yolk sac tumour (YST) is the most common histological subtype of MGCT and usually involves Gonads (testes and ovaries). Affected individual are almost exclusively infants, and all are younger than three year of age at presentation.1 Extra-pineal gonadal site are rare and involves mediastinum, endometrium, cervix, vagina, gland and sacrococcygeal area.1 Early integration of palliative care with standard oncology care provides adequate symptoms control and total care of patients and caregivers.²⁻⁴ Paediatric Palliative care differs from adult palliative care and requires special attention to physical, developmental, psychosocial, ethical, spiritual issues that are unique to each child. In this study we describe a case of rare site uterus and vagina YST in one-year old child with vaginal bleeding causing psychosocial distress in family. A child's life-threatening illness profoundly impacts family both emotionally and financially. Early palliative care in paediatric patients reduces the suffering of child and parents.

Case report

A one-year old female patient was brought in January 2018 with history of vaginal bleeding for twenty-five days. Radiological studies (USG and MRI) showing well defined lesion involving uterus and vagina. Elevated level of serum alfa fetoprotein, normal B-human chorionic gonadotropin and

Immunohistochemical studies favoured the diagnosis of Yolk Sac Tumour. The patient received three cycles of JEB regime (Carboplatin + Etoposide and Bleomycin) from 9th January to 19th February 2018 which resulted in radiological resolution of lesion and decreased AFP level (19.39ng/ml). After four months, there was recurrence of lesion posterior to urinary bladder at upper vagina with lung metastasis and raised AFP (625.50ng/ml) for which patient received six cycle of VeIP regime (Cisplatin + Ifosfamide + Vinblastin) from 24th July 2018 to 21st February 2019 resulting in resolution of both vaginal and lung metastatic lesion. Patient received six cycle of TIP regime (Paclitaxel + ifosfamide + Cisplatin) from 6th June to 26th August 2019 for recurrent lesion at vaginal wall. Further chemotherapy was planned, but patient lost for follow up. After five-months patient was brought with vaginal bleeding and radiological studies showed tumour metastasized to lung and liver. Patient was planned for palliative radiotherapy to primary site but parents refused. At this point they were referred to Department of Palliative Medicine.

When they visited palliative medicine department...

While parents brought the child to our OPD, she was continuously, inconsolably crying. On quick examination, she was found having abdominal pain (Wong baker faces scale 9/10). On history taking, her condition was described same for one week. She was on syrup paracetamol 12mg/kg three times a day. She had history vaginal bleeding and was on Tab Tranexamic acid 20 mg/kg in three divided doses a day. They were coming for treatment from another state, 1000 kms away. She had two elder sisters staying with her grandparents. During communication with parents, we found that parents were educated, social, spiritual, co-operative, and economically belonged to middle class.

Parents were emotionally drained due to unacceptable physical suffering of their child, psychologically stressed due to worries of other daughters being left alone to be cared by grandparents. Economically, also it was burdensome to travel frequently or to stay in vicinity of hospital for long-time. They were confused and in spiritual dilemma about refusing the radiotherapy, an unfruitful treatment at this stage.

Patient was quiet and comfortable after receiving 1mg Morphine IR orally. Tab Morphine IR 10 mg was dissolved in 10ml purified water and precise 1ml was given orally by syring. We have followed this practice of Morphine administration as Morphine suspension is not commercially available. Parents were taught to prepare 1 mg oral morphine in similar way at home. They were advised to give 1ml every four hours and half ml for breakthrough pain if required, along with other supportive treatment.

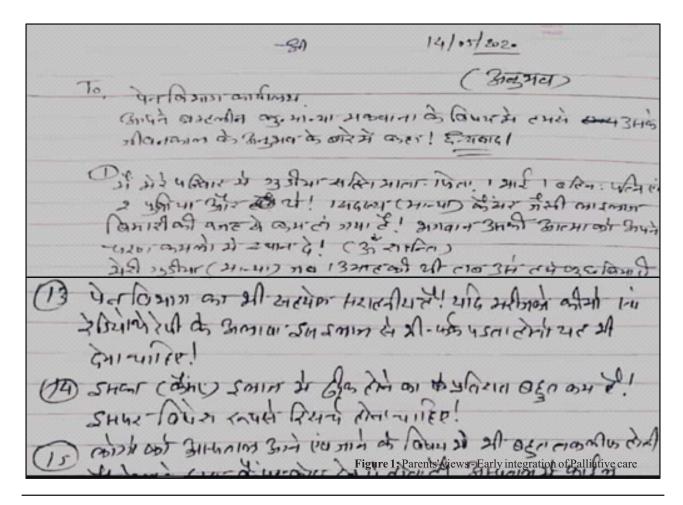
Parents were counselled for disease status and prognosis, round the clock use of medications, end of life care, general nursing care and place of care preference (hospital, hospice or home). All their questions were answered empathetically. Parents chose home care and took the child home to her sisters and grandparents with satisfaction of receiving proper guidance and relief from guilt. Palliative care team remained in constant contact with them through phone calls, and found that she expired peacefully after two weeks.

Discussion

Vaginal YST are rare and clinically present with history of bloody vaginal discharge which is often accompanied by polypoid mass protruding from the vagina. Vaginal YST are both locally aggressive and capable of metastasis via hematogenous and lymphatic pathway. The serum AFP level remains a useful marker for diagnosis and monitoring the recurrence of vaginal YST in infants. Combined chemotherapy and surgical resection remain the mainstay of treatment. Patients and caregivers are not routinely screened for psychosocial-emotional distress.⁶ Also paediatric patients are not prescribed strong analgesic for optimum pain relief because of lack of knowledge regarding its use in children.^{6,7} Paediatric palliative care is also less utilised in our country and still palliative care references are done late in the disease trajectory.8

In paediatric palliative care¹⁰ the transition from health to ill health may occur in four distinct ways.

- 1. Have a potentially curable illness but treatment fail
- 2. May receive intensive treatment that can be expected to prolong the life but child is likely to die before adulthood



- 3. Are diagnosed with progressive condition for which no curative treatment exists
- 4. Have a progressive condition but are vulnerable to early death as a result of general debility and morbidity such as respiratory infection.

In our case, YST falls in category one and received intensive treatment. For most children with malignancy cure is probability or possibility at the time of diagnosis. At that time child and family need support in living with uncertainty. This is called upstream palliative care in which the seriousness of condition is revisited and discussed at regular interval during the child's illness. Paediatric patients and their families make multiple transitions during the course of illness. The goal of care should be discussed regularly and renegotiated so that they reflect on the child's care in changing circumstances. This can be possible only if palliative care is integrated early.

In this case, due to pain and symptoms burden of the child, parents had lots of suffering throughout the disease trajectory, which includes emotional distress, lack of communication, financial issues, transportation problem etc. Early palliative care improves quality of life by adequate symptom relief, good emotional, spiritual, and psychosocial support to the patients and caregivers.^{2,3}

Conclusion

In this case, though patient received only one palliative care consultation during the last visit to hospital, parents appreciated adequate control of physical symptoms, pain management and guidance in decision making by giving realistic prognostic information and emotional support in end of life care (Figure 1). In India where parents remain the sole decision makers for the child, it is utmost importance for bridging the gap in cancer care by providing both cancer treatment and psychosocial-spiritual support to parents in paediatric patients.

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