Case Report-III

Extragonadal Germ-Cell Tumour of Vagina.

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ABSTRACT:

A 7-month-old female child was admitted with a short history of bleeding and mass protruding from the vagina. Alfa-feto protein levels were raised. Biopsy taken of the tumour mass was suggestive of Endodermal sinus tumour (yolk sac) of vagina. Neoadjuvant chemotherapy was given. Organ preserving resection of tumour was done successfully.

INTRODUCTION

Malignant germ-cell tumour (MGCT) is a rare tumour of childhood accounting for less than 3% of paediatric malignancies. The vagina is extremely rare site for germ cell tumours. Endodermal sinus tumour forms the most common histological subtype of MGCT.

CASE : A 7-month-old female child was admitted with history of bleeding per vagina. Child was pale and had tachycardia. Clinical examination revealed mass protruding from vagina (fig 1). Serum Alpha feto protein level AFP was 664ng/dl (Normal range for this age 9.7+/- 7.1). CT scan of pelvis showed the tumour arising from posterior wall of vagina sized 1.2 X 1.5 Cms (fig 2). Biopsy was taken per vaginum. Histopathological examination of tumour mass was suggestive of yolk sac tumour (highly malignant variety). Neo-adjuvant chemotherapy using BEP [Bleomycin (B), Etoposide (E), and Cisplatin (P) at an interval of three weeks was given. At the end of third cycle, serum AFP levels dropped to

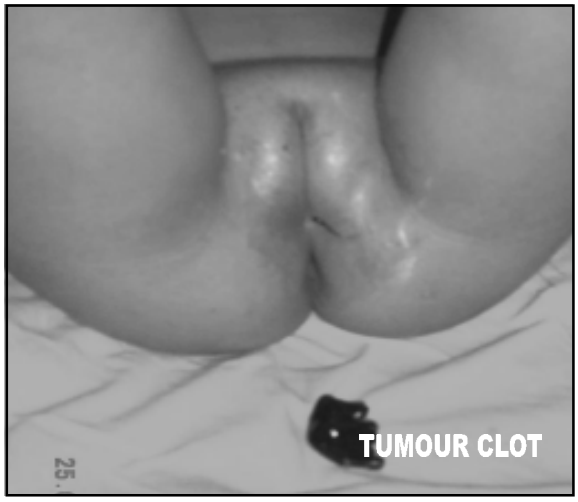

Figure1: Clinical photograph showing tumour mass per vagina.

8ng/dl. Repeat CT scan showed a residual tumour of 0.9 x 0.6 Cms. Surgical excision of the residual tumour with preservation of vagina was done. The histopathological examination was suggestive of fibrotic nodule with no residual tumour. Two more cycles of Etoposide (E) and Cisplatin(P) were given; following which the AFP levels were normal. There was no evidence of recurrence after 6 months of follow-up.

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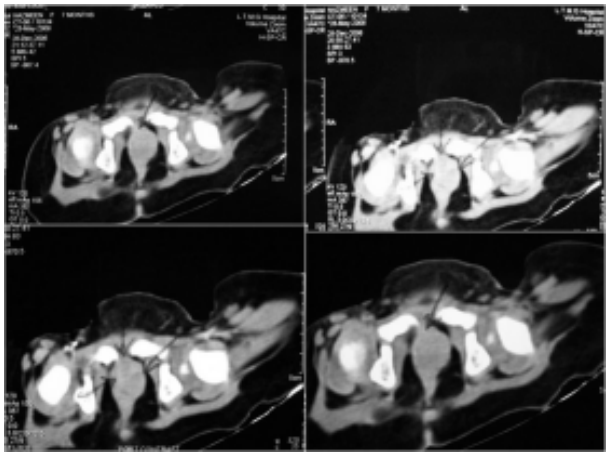

Figure 2: CT scan showing a 1.2 X 1.5 cms tumour arising from posterior wall of vagina.

DISCUSSION:

Endodermal sinus tumour was first described by Telium in 1959.[1] Malignant germ-cell tumours (MGCT) of childhood account for less than 3% of paediatric malignancies.[2-7] The vagina is extremely rare site for EST and is usually diagnosed before 3 years of age.[1,2,7] Literature revealed only about 50 cases so far. Rare sites are ovary, pleura-pulmonary, testes, head, chest, abdomen, pelvis, and sacrococcygeal (lower back) area.[2] Presentation is mostly-bleeding and mass protruding through vagina. Minimally invasive surgery with neo-adjuvant chemotherapy is the treatment of choice. The histiogenesis of this rare site may be due to aberrant migration of germ cells during early embryonic life from the yolk sac to the gonadal ridges.[2]

The clinical presentation of this tumour includes history of bloody or blood tinged vaginal discharge, sometimes accompanied by a polypoidal mass protruding from the vagina. Most common differential diagnosis is botyroides sarcoma differentiated by its characteristic grape like appearance and histopathology.[2]

Radiological imaging by CT or MRI scan may demonstrate the tumour extension into the surrounding structures.[1,2,7] The histological diagnosis is based on the pathognomonic evidence of Schiller-Duval bodies.[1] Serum AFP levels can aid in pre-operative diagnosis, monitor effectiveness of therapy and detect recurrence before clinical manifestations.

Management of this vaginal tumour should take into consideration the chemo-sensitivity of tumour and preservation of organ and its function. Endodermal sinus tumour is chemo sensitive.[1] In our case tumour size was reduced by almost 50% and was successfully removed with preservation of vagina. On histopathology residual mass showed fibrotic nodule with no malignant cells. Tumour sensitivity to chemotherapy has made it possible to give neo-adjuvant chemotherapy thus minimizing surgical excursion. This allowed preservation of sexual function in this case. Delayed surgical resection with organ preservation is not associated with an adverse outcome. The current survival rate for genital MGCT is excellent. The 4-year event-free survival rate was 76.2% ± 13.1%, and 4-year overall survival rate was 91.7% ± 8.4%in one study.[8]

We emphasize that neo-adjuvant chemotherapy may be used with minimal surgical excursion for EST of vagina to allow preservation of sexual function.

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