http://jmscr.igmpublication.org/home/
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v8i8.04

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Rhabdomyosarcoma of the vagina in a 30-month-old girl: Refractory to conventional chemotherapy

Authors
Faisal R GURU[1], Shah Saqib Ahmad[1], Aiffa Aiman[2], Javvid Muzammil[3], Gul Muhammad[1]
[1]Department of Medical Oncology and Stem cell transplant SKIMS Soura
[2]Department of Pathology SKIMS Soura
[3]Department of Medical Oncology GMC Srinagar, India

Introduction

Gynecologic tumors in children are rare and represent <5% of all pediatrics neoplasms(1). Rhabdomyosarcoma (RMS) are malignant mesenchymal tumors originating from myogenic progenitor cells. They represent the most common soft tissue tumor in childhood. The head and neck region are the most affected tumor sites, followed by the genitourinary tract(2). Four major histologic subtypes of RMS are identified, Embryonal, Alveolar, Botryoid, Pleomorphic/ undifferentiated (2,3). Botryoid variant is a type of embryonal RMS arising within the wall of the bladder or vagina, can also occur in the cervix. This tumor is seen almost exclusively in infants, it is characterized by 'grape-like' appearance caused by polypoid mass arising in submucosal tissue(2,4). Embryonal RMS of the vagina is a variant which represent an extreme rare and aggressive form associated with poor prognosis(2)

Case Report

An 30-month-female, full term, presented with history of vaginal swelling at the age of 28 months. A red small mass protrude through the introitus was noticed at the age of 28 months by the mother initially; there was also difficult micturition without change in urine's color or odor. No history of medication use by mother during pregnancy. Family history was unremarkable

The baby appeared well, vitals stable.Abdomen was distended with no palpable inguinal lymph node, no hepato-splenomegaly. Pelvic examination revealed a rounded red mass 5 × 5 cm, protruding through the vaginal introitus, normal labia minora and majora. Digital rectal exam: palpable non-fixed mass pushing on the rectum. (Figure1) Other examinations were within normal.

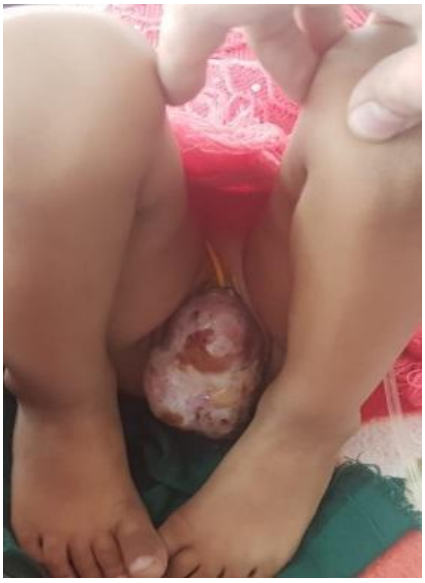

Figure 1

Ultrasound abdomen showed heterogeneous mass arising from the posterior wall of the vagina measuring 31*38*38mm. (Figure2) Contrast enhanced CT abdomen showed a 5*4.9*5.8cm heterogeneous mass in the recto uterine pouch abutting and displacing the rectum and uterus. Bowel loops were displaced but no sign of intestinal obstruction/invasion, liver, spleen, lung and mediastinum were normal. (Figure 3)

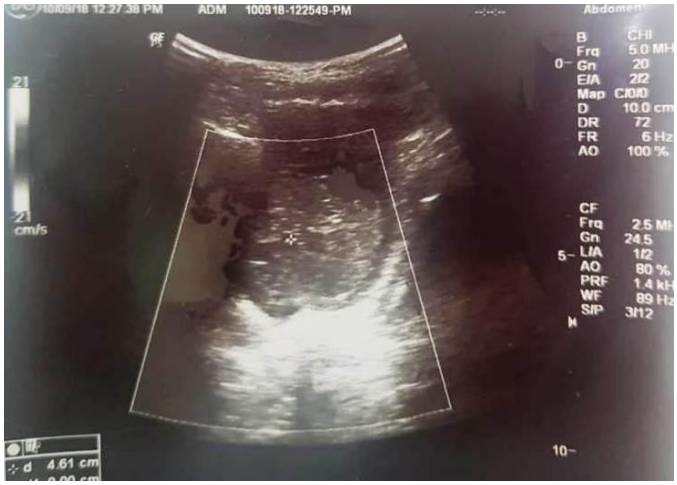

Figure 2

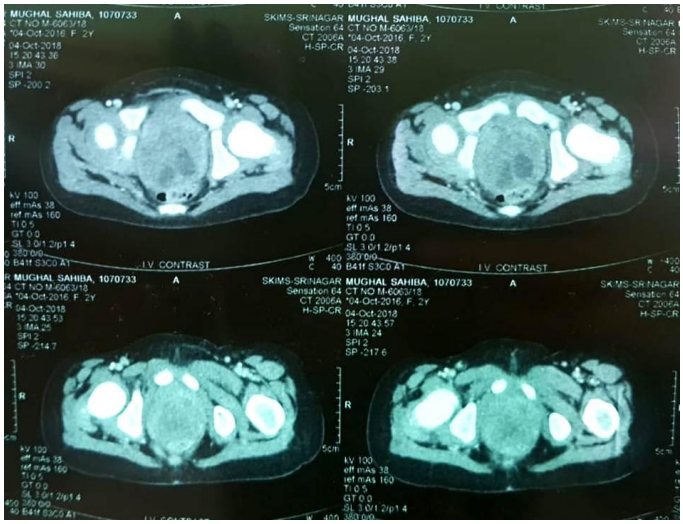

Figure 3

The child underwent a cystoscopy with debulking of tumor. The cystoscopy findings revealed a grossly normal bladder with a mass 5*5cm arising from the posterior vagina. A biopsy was taken and histopathology was consistent with embryonal Rhabdomyosarcoma WHO Grade 3 tumor showing a differentiation score of 3, mitotic count greater than 20 per 10 HPF, degree of necrosis less than 50%. Desmin, Myogenin, CD 99 were positive and S-100, SMA was negative. (Figure 4)

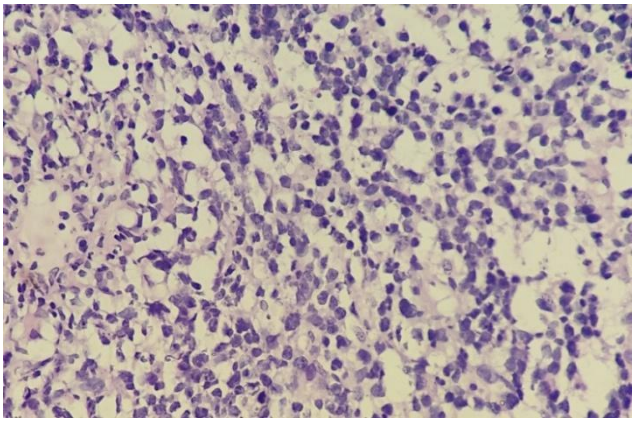

Figure 4

Bone marrow Aspiration and Biospy was normal. The patient received 3 cycles of vincristine, actinomycin D and cyclophosphamide but there was progression of disease. Re assessment was done and USG revealed a 10 * 8 cm mass arising from the posterior vaginal wall. The chemotherapy was changed to ifosphomide plus etoposide and the patient responded almost 50%

reduction in size after 2 cycles. The child is planned for reassessment and if surgically resectable will undergo surgery followed by more chemotherapy.

Reference

1. Fernandez-Pineda I, Spunt SL, Parida L, Krasin MJ, Davidoff AM, Rao BN. Vaginal tumors in childhood: the experience of St. Jude Children's Research Hospital. J Pediatr Surg2011;46:2071–5.
2. Hartmann A, Thiel FC, Beckmann MW, Lux MP. 18-year-old woman with an embryonal rhabdomyosarcoma of the uterus in statu nascendi. Geburtshilfe Frauenheilkd2012;72:1132–6.
3. Agarwala S. Pediatric rhabdomyosarcomas and nonrhabdomyosarcoma soft tissue sarcoma. J Indian Assoc Pediatr Surg 2006
4. Akhavan S. Sarcoma botryoides (embryonalrhabdomyosarcoma)o f the uterine cervix in sisters. J Gynecol Oncol 2010;21:273–5.