

Case Report

A Case Report of Rhabdomyosarcoma of Uterine Cervix in a 7-Month-Old Child

Yasmin F^a, Ahmed MAU^b, Begum T^c, Ahmed T^d, Baki MA^e

Abstract

Rhabdomyosarcoma is the soft tissue tumor derived from embryonic primary mesenchyme. Neck, head and genitourinary region are the common locations of rhabdomyosarcoma.

Here, we report a 7-month-old child who presented with a protruding mass per vaginally. Abdominal examination revealed a 6 cm x 6 cm hard, irregular, non-tender mass at pelvic region. Ultrasound of abdomen showed large heterogenous pelvic mass of uterine origin. Magnetic resonance imaging of whole abdomen revealed an intrapelvic teratoma (9.5 x 7.4 x 10 cm³) with left sided hydronephrotic change as the mass was pressing the rectum and bladder. The child underwent a total hysterectomy. Histopathology result was suggestive of rhabdomyosarcoma of uterus.

Keywords: Rhabdomyosarcoma, Hysterectomy, Cancer.

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Introduction

Rhabdomyosarcoma (RMS) is the most prevalent malignant soft tissue tumor among children. The embryonal subtype is more common during the first decade of life, whereas alveolar RMS (ARMS) frequently follow in 10 to 25 years of age.¹ Only 6% of all RMS cases are found in infancy.^{2,3} RMS has a special predilection for head and neck region, genitourinary tract and extremities.⁴

We present here an unusual case of a large intra-abdominal embryonal RMS.

Case Report

A 7-month-old female infant was brought by her parents to the Pediatric Outpatient Department of Bangladesh

Institute of Research and Rehabilitation for Diabetes, Endocrine and Metabolic Disorders General Hospital in 2015 with protruding mass in the vaginal area for 7 days and crying during micturition for same duration. Abdominal examination revealed a 6 cm x 6 cm² hard, irregular, non-tender mass at pelvic region. Skin over the mass was normal. Size of the protruding per vaginal mass was 2.5x 3 cm², which was irregular in outline, pinkish in color with no contact bleeding (Fig. 1).



Figure. 1 Protruding per vaginal mass

Author Information

- Dr. Farzana Yasmin, Resident Medical Officer, Dept. of Paediatrics, BIRDEM General Hospital.
- Dr. Md. Ashraf Uddin Ahmed, Resident Physician and Jr. Consultant, Dept. of Medicine, BIRDEM General Hospital
- Dr. Tahmina Begum, Professor, Dept. of Paediatrics, BIRDEM General Hospital.
- Dr. Tasnima Ahmed, Registrar, Dept. of Paediatrics, BIRDEM General Hospital.
- Dr. Md. Abdul Baki, Asst. Professor, Dept. of Paediatrics, BIRDEM General Hospital.

Address of correspondence: Dr. Farzana Yasmin, Department of Paediatrics, BIRDEM General Hospital 2, Dhaka, Bangladesh. Email: shakilbm14@yahoo.com

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Ultra sonogram of abdomen showed large well defined piriform shaped soft tissue echogenic mass lesion in pelvic cavity measuring 12 x 6.8 x 7.1 cm³ extending

up to umbilical region. Echo texture was heterogeneous, on color flow, marked vascularity was noted. Magnetic resonance imaging (MRI) of whole abdomen revealed a fairly large lobulated mass having intimal septation and measuring about $9.5 \times 7.4 \times 10 \text{ cm}^3$ in the pelvic cavity displacing the adjacent bowel loops and compressing the left ureter causing left sided hydronephrosis. On T1 weighted film the mass was predominantly isointense with hypointense component while on T2 weighted film the mass was heterogeneously hyperintense with hypointense component. Following contrast augmentation heterogenous enhancement of the lesion was evident (Fig. 2). Malignancy screening showed raised level of carcinoembryonic antigen (CEA) 19.32 ng/mL (Reference value: 0-5.0 ng/mL), alpha feto protein (AFP) 7.74 ng/mL (Reference value: <13.46 ng/mL) and cancer antigen 125 (CA-125) 379.10 U/mL (Reference value :< 35 U/mL).

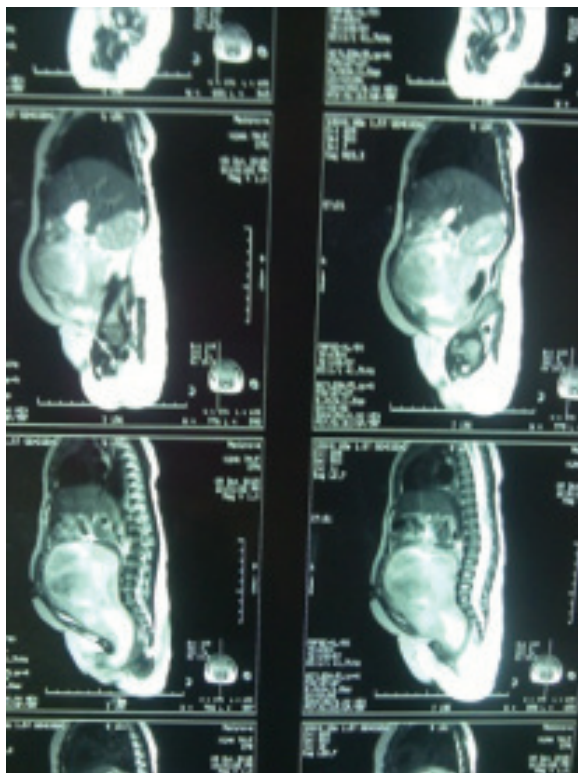


Figure. 2 T2 weighted sequence of MRI showing contrast of the mixed intensity lesion

The patient underwent subtotal hysterectomy and tissue was sent for histopathology. Her histopathology report yielded a malignant tumour made up of spindle shape

cell with equivocal distribution of undifferentiated and well-differentiated rhabdomyoblasts in absolute absence of any pleomorphic rhabdomyoblasts. She was advised to take 14 cycles chemotherapy by oncologist. But unfortunately after receiving 5 cycles, parents discontinued the therapy and switched to homeopathic medication for 2 months. Subsequently abdominal mass reappeared for which they consulted with surgeon and resection of the mass was done again. Now she is on chemotherapy.

Discussion

RMS is a common neoplasm, representing 5 - 10% of malignant solid tumors in childhood and is the commonest soft-tissue sarcoma in the paediatric age group.⁵

The two main histological subtypes of RMS found in the paediatric age group are embryonal (with botryoid and spindle cell variants) and alveolar. The remaining cases are of pleomorphic or undifferentiated.⁶ The aspirates in present case yielded a malignant tumor made up of spindle shape cell with equivocal distribution of undifferentiated and well-differentiated rhabdomyoblasts in absolute absence of any pleomorphic rhabdomyoblasts. Spindle cell RMS, as described in our patient, is a rare variant of the embryonal type with a favorable clinical course. This tumor tends to arise in younger patients, with a predilection for males, which is not similar to our case.

MRI is the best imaging modality in RMS due to its excellent ability to determine soft tissue changes. Computerized tomography (CT) scan is useful in assessing any metastatic manifestation from RMS.⁷ Considering the reported case, MRI confirmed the abdominal wall constituents as origin of the mass, ruling out any visceral involvement. Corresponding CT scan further excluded any visceral metastasis.

Because of the varying cellular distribution in different regions of a RMS mass, the pleomorphic rhabdomyoblasts from anaplastic RMS often fail to be represented in the aspirated smears.⁸ Recapitulating similar phenomenon, the aspirates in present case yielded equivocal distribution of undifferentiated and well-differentiated rhabdomyoblasts in absolute absence of any pleomorphic rhabdomyoblasts. Under these cytomorphological considerations the mass was diagnosed as embryonal RMS (ERMS); however,

histopathology revealed the presence of pleomorphic multinucleated rhabdomyoblasts in conjugation to classic ERMS morphology.

Finally, the resected mass from the reported case was rendered the diagnosis of anaplastic RMS. The presence of characteristic well-differentiated rhabdomyoblasts, in examined smears and sections, clinched the diagnosis of RMS in this case, nullifying the necessity of any additional diagnostic work-ups.

The diffuse distribution of well-defined rhabdomyoblasts in the discussed neoplasm readily depicted its rhabdomyoblastic phenomena, dismissing the need of any further ancillary tests, and virtually ruled out the possibility of any divergent neoplasms.

Depending upon the size of tumor, various treatment modalities have been described as combined brachytherapy and chemotherapy without surgery, only surgery, chemotherapy with surgery and triple therapy comprising of chemotherapy, and surgery as well as radiotherapy.⁹ Peripheral stem cell support is associated with longest survival among RMS patients.¹⁰

Conflict of interest: None

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