A 17-YEAR-OLD MALE WITH PROSTATIC RHABDOMYOSARCOMA

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Abstract:

Prostatic rhabdomyosarcoma is a common tumour in infancy and childhood but rare in young adults and older people. A 17-year-old boy presented with features of bladder outlet obstruction. On digital rectal examination, a growth was found in the prostate. After performing trans-rectal ultrasound guided biopsy it revealed rhabdomyosarcoma of prostate which was locally advanced, involving bladder pelvic nodes. Neoadjuvant chemotherapy with ifosfomide and doxorubicin was given for 6 cycles. However, it was still inoperable and therefore, pelvic radiotherapy was started improving the local control.

Key words: *Rhabdomyosarcoma, prostate, soft tissue, sarcoma.*

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Introduction:

Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in the paediatric age group constituting 3-4% of all malignancies. Approximately 350 new cases are reported with RMS below 20 years of age in the United States.[1] It is more common in males than female and most cases occur under 10 years of age. The median age at diagnosis is 5 years.[2] Although cases are sporadic, some genetic and environmental factors have been associated with this type of cancer like germline mutation, Costello syndrome, Beckwith Widemann syndrome, Neurofibromatosis type I, paternal use of cocaine and marijuana, birthing order and accelerated growth in utero.

RMS can occur in any part of the body, but the most frequent sites are the head and neck region (35%) followed by the genitourinary tract including prostate, urinary bladder, vagina, vulva, uterus and paratesticular area (26%) and the extremities (19%). The International Classification of Rhabdomyosarcoma recognizes three main categories depending on their outcome:

1. Superior prognosis
   a. Botryoid
   b. Spindle cell
2. Intermediate prognosis
   Embryonal RMS
3. Poorer prognosis
   a. Alveolar RMS
   b. Undifferentiated sarcoma

In paediatric age group the embryonal rhabdomyosarcoma (ERMS) (65%) and alveolar type of RMS (25%) are most common.[3] Clinical presentation depends on the location of the primary tumour and spread of the disease. The head neck tumours may present with proptosis, ophthalmoplegia, nasal discharge, nasal obstruction, headache, cranial nerve palsy, dysphonia, dysphagia and palpable adenopathy.[4] The patients with genitourinary RMS may present with dysuria, haematuria, hydronephrosis, palpable abdominal mass, vaginal discharge. The extremity RMS may present with palpable mass, pain or palpable lymphadenopathy.[4] The staging evaluation should

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include complete blood count, blood chemistries, bone scan, Positron Emission Tomography (PET) scan, bilateral bone marrow aspiration and biopsy, Computed Tomography (CT)/Magnetic Resonance Imaging (MRI) of the primary tumour site, CT scan of chest, CT/MRI of abdomen and pelvis. Sentinel node biopsy is gradually increasingly used for assessment of the extremity tumour. Clinical group, stage, histologic subtypes, age, treatment are important prognostic factors.[5] Management of RMS needs multidisciplinary team; surgery, chemotherapy, radiotherapy and biologic therapy all have role to manage this malignancy.

Case Summary:

A 17-year-old male admitted in the department of Radiotherapy, Dhaka Medical College Hospital on 18 February, 2017 with the complaints of difficulty in micturition for 10 months. He had increased frequency and urgency of micturition which worsened at night. He also felt hesitancy and pain during micturition. The boy needed to strain to initiate and maintain urination. He also had reduced force of stream as well as feeling of incomplete evacuation and dribbling. For the last two months he developed lower abdominal pain, which was continuous and dull aching in nature, sometimes radiating to back. Pain worsened during voiding urine and was not relieved by any medication. He also developed nausea and vomiting for last two months. Initially he vomited 1-2 times a day but frequency of vomiting gradually increased to even 6-7 times a day. Vomiting was non-projectile, amount of vomitus varied from small to profuse and there was no diurnal variation. For last ten days the boy complained of inability to void urine and gradual fullness of lower abdomen. He also developed generalized swelling of the body specially face and legs for last seven days. He gave no history of haematuria, fever, weight loss, cough, bony pain or contact with tuberculosis patient. His bowel habit was normal. He is nonsmoker, non-alcoholic, normotensive and non-diabetic.

Perurethral catheterization was done to relieve urinary retention at local hospital and he was referred to the department of Nephrology, DMCH for haemodialysis. Then the patient was referred to the urologists for DJ stenting and ante-grade DJ stent was placed. Then he was referred to the department of Radiotherapy, DMCH for further management.

On examination, he was ill looking with puffy face, moderately anaemic and oedematous. There were drain tubes in both flanks and a per-urethral catheter in situ. Incisional scar marks were present in both inguinal regions. He was haemodynamically stable and afebrile. On digital rectal examination, rectal mucosa was smooth and there was a non-tender growth at prostatic area, firm to hard in consistency and irregular in shape. Rectal wall could not be moved freely over the growth.

On investigation, his haemoglobin concentration was 8.5 gm/dl and serum creatinine was 12.1 mg/dl. Other blood chemistries were normal. After three sessions of haemodialysis his serum creatinine became normal. His serum prostate specific antigen (PSA) level was 0.97 ng/ml which was normal. Ultrasonogram of whole abdomen revealed right sided pelvic mass involving lower right ureter, mild hydroureterocele and mild splenomegaly.

CT scan of Whole Abdomen showed a heterogenous enhancing mass lesion arising in the prostate with possibility of extracapsular extension, involving the urinary bladder wall and right distal ureter with few locoregional enlarged lymph nodes, mild hydronephrosis and Splenomegaly (Fig. 1). Magnetic resonant urography (MRU) showed mildly dilated pelvicaliceal system with narrowing at pelvi-ureteric junction (PUJ) with mild hydronephrosis in right kidney (Fig. 2).

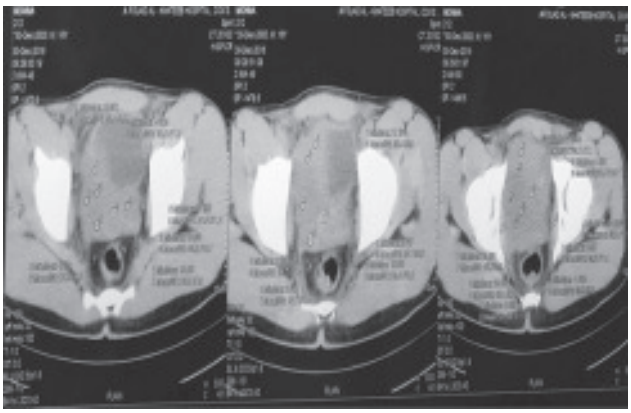


Fig.-1: *CT scan of abdomen showed a heterogenous enhancing mass lesion arising in the prostate*

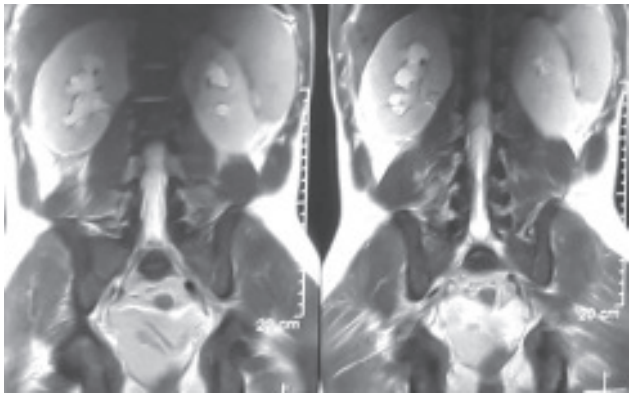
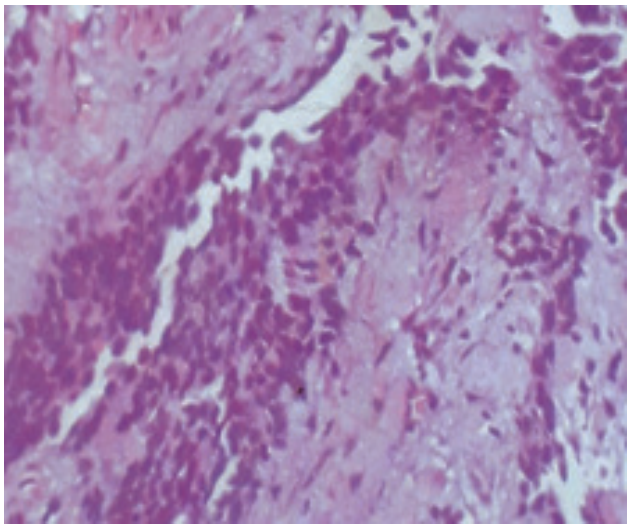


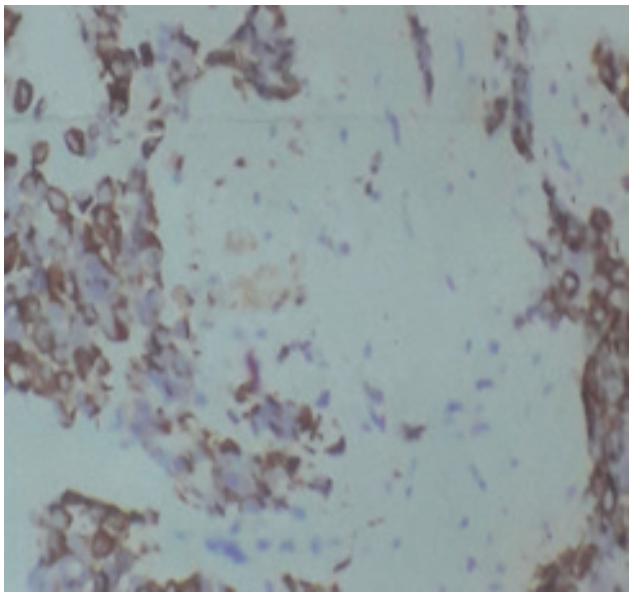
Fig.-2: *MR Urography showed mildly dilated pelvicaliceal system with narrowing at pelvi-ureteric junction (PUJ) with mild hydronephrosis in right kidney.*

Transrectal prostatic tissue biopsy from right lobe revealed RMS (Fig. 3). Prostatic tissue taken from left lobe of prostate showed benign prostatic tissue. Immunocyto-histochemistry (IHC) was compatible with RMS (Fig. 4).



H&E Rhabdomyosarcoma, D/D-primitive neuroectodermal tumour

Fig.-3: *H&E staining of prostatic tissue showing features compatible with rhabdomyosarcoma*



Desmin; positive

Fig.-4: *Immunohistochemistry of prostatic tissue*

After confirmation of the diagnosis, systemic chemotherapy was started with ifosfamide and doxorubicin 3 weekly. The patient was reviewed for surgery from the department of Urology after 6 cycles of chemotherapy. He was declared inoperable and was then planned for radiotherapy to the pelvis and prostatic bed with 6 MV LINAC at 200 cGy per fraction with total dose up to 60 Gy.

Discussion:

Malignant prostatic tumours of mesenchymal origin are rare, corresponding to 0.3% to 1.0% of all prostatic tumours. Thirty percent of these tumours are ERMS.[6] Although primary RMS of prostate is common in paediatric age group, it is rare in adults.[7] The natural history of prostatic RMS is characterized by rapid growth with the formation of pelvic or abdominal masses, renal failure due to uropathy. This type of tumour also spreads to distant sites like lungs, bone, liver and also to the regional lymph nodes. The presenting symptoms are usually due to obstruction of the urethra like frequency, hesitancy, dysuria and less commonly haematuria and acute urinary retention. It may cause constipation, rectal fullness or rectal bleeding due to compression of the rectum. There is no pathognomonic radiologic finding for RMS. CT scan or MRI study can reveal a large soft tissue mass with areas of necrosis in the prostate. The mass may invade peri-urethral and perivesical soft tissues even up to ishchiorectal fossa. The role of PET-CT scan for staging is yet to be evaluated but in some reports it is useful in the detection of the metastatic disease or obscure metastasis. The diagnosis is confirmed by histopathology of the biopsy specimen taken (perurethral, transrectal or perineal biopsy). The histopathology report can identify different grades. In most of the cases IHC for skeletal muscle specific markers such as actin, myosin, desmin can confirm the tumour as RMS.

The treatment and prognosis of the urogenital RMS in children have changed now-a-days. Now with the aid of chemotherapy, precise radiation and even biologic therapies help to achieve remarkable cure rate with good quality of life. But in adults, RMS of the prostate is very rare tumour with poor prognosis. Most of the patients present at advanced stage due to rapid growth and the absence of early symptoms. One-fourt of the patients may have distant metastasis at the time of initial presentation. Treatment options depend on the stage at the time of diagnosis. As most of the cases are presented with advance stage, treatment mainly starts with neoadjuvant chemo-radiation therapy. In the metastatic disease, the role of local treatment with radiation is promising. It can improve progression free survival (PFS) and also overall survival (OS). However, the timing of each modality of the treatment

must be individualized depending on condition and symptoms of the patient.

In a retrospective review of 48 patients with urological sarcoma by Lee *et al.*[7] it was shown that any local treatment can improve survival of the patients irrespective of the distant spread of the disease. In adults, prognosis of the prostatic RMS is poor with median survival of 16 months from the time of diagnosis; most of them died due to metastatic disease. Primary embryonal rhabdosarcoma of the prostate gland is an extremely rare pathological and clinical entity in adults[8], with fewer than twelve cases reported in the literatures[9]. The prognosis is poor, with a five-year survival rate of 30%-35% for all RMS in adult individuals[10]; this rate is probably lower in cases of primary tumor on the prostatic bed.[11]

Conclusion:

RMS of the prostate is a very rare tumour in adult population and presents in advanced stage. Optimum therapeutic approach still is not defined due to small number of cases. Therefore, for early diagnosis strong clinical suspicion is needed in any adult person presenting with symptoms of bladder outlet obstruction (BOO) resulting in better clinical outcome.

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