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Paediatric Surgery

RAPIDLY PROGRESSING PELVIC LUMP: WORKUP OR RESECTION TO PREVENT IMMINENT ABDOMINAL COMPARTMENT SYNDROME.

Dipanjan Goswami	Department of Paediatric Surgery, Maulana Azad Medical College.
Sheetal Upreti*	Department of Paediatric Surgery, Maulana Azad Medical College. *Corresponding Author
Gaurav Saxena	Department of Paediatric Surgery, Maulana Azad Medical College.
Sujoy Neogi	Department of Paediatric Surgery, Maulana Azad Medical College.
Simmi K Ratan	Department of Paediatric Surgery, Maulana Azad Medical College.

ABSTRACT Rhabdomyoscarcoma of bladder in paediatric population is a rare tumour with guarded prognosis. Upfront surgery is indicated only in cases of possibility of complete resection followed by chemotherapy and radiotherapy. We present a case of rapidly progressing lump with imminent abdominal compartment syndrome, for which upfront resection was done to alleviate patient symptoms followed by chemotherapy and radiotherapy.

KEYWORDS : Bladder RMS, Rhabdomyosarcoma, Pelvic mass

INTRODUCTION:

Pelvic masses in female can range from ovarian masses, neuroblastoma to rhabdomyosarcoma (RMS). RMS stands out as the predominant soft tissue sarcoma found in paediatric and adolescent demographics.[1] It is the third most common extracranial solid tumour of childhood after Wilm's tumour and Neuroblastoma. RMS arise from mesenchymal tissues, allowing them to manifest in various parts of the body. While they can develop almost anywhere, the trunk, extremities, and head/neck areas are frequently impacted. Notably, around 18–22% of cases occur in the genitourinary tract establishing it as the second most prevalent location for RMS.[2] Our case was a rapidly growing lump originating from the pelvis and occupying almost the whole of the abdomen in a month's time. Onset of abdominal compartment was imminent considering the rate of the growth of the lump. The purpose of the case report is to bring forth the diagnostic dilemma with regards to the origin of the lump and also the choice of treatment to follow. On one hand, the conventional approach is to do workup of the tumor, while on the other hand upfront resection considering the rate of the growth of the tumor.

CASE SUMMARY:

7 year old girl, presented with incidentally detected lower abdominal mass noticed by the mother, and the mass rapidly progressed to occupy almost 90% of the abdomen, in one month. There was no other significant associated history. On examination there was a lump of approximately 10 x 12 x 12 cm arising from pelvis, occupying almost whole of the abdomen except left hypochondrium and a part of the epigastrium (Figure1). On investigations, the tumour markers (AFP and Beta hCG) were within normal range. CT revealed a large well defined intraperitoneal predominantly solid mass with multiple cystic areas with lobulated contours (10 x 13 x12 cm).(Figure2) The small bowel loops were displaced superiorly and to the left side of abdomen. Uterus was normal. Right ovary was not seen separately. Left ovary appeared normal. There were nodules in the left upper lobe which was reported as metastasis. Patient underwent urgent cystoscopy followed by exploratory laparotomy as the tumour was rapidly expanding and spreading locally. On cystoscopy there was external mass effect on the dome of bladder. No extension of growth inside the lumen of bladder was noticed. On Exploratory Laparotomy, a large mass was present in lower abdomen arising from dome of bladder (Figure3). Consistency was variegated. Omentum was densely adherent over tumour surface, bowel was pushed superiorly and uninvolved. Left lateral pelvic wall was free of adhesion. There were no peritoneal tumour or liver deposits. Bilateral ovaries were normal. While mobilizing and dissecting the mass, there was intra-peritoneal rupture of tumour capsule with spillage. While resecting from bladder, the right lateral part of dome of bladder was resected. Lymph node dissection done along the right Iliac vessels. Bladder rent was repaired in 2 layers. Titanium clip was applied at the site of repair of bladder wall and right lateral pelvic wall to facilitate targeted radiotherapy. The post-operative course was uneventful. The biopsy proved the tumor to be Bladder Embryonal Rhabdomyosarcoma with omental and retroperitoneal deposits. PET CT was done to delineate lung nodules and it showed that they were malignant. Child was started on Vincristine, Actinomycin D and Cyclophosphamide (VAC regimen) on post-operative 7th day and planned for radiotherapy. After completion of 2 cycles (week 0,1,2,3,4,5) of VAC chemotherapy, patient underwent whole body PET scan –which revealed few small non FDG avid bilateral lung metastases. As compared to previous CT scan, abdominal and pelvic soft tissue mass was no longer visualised and bilateral lung lesions showed decrease in size.

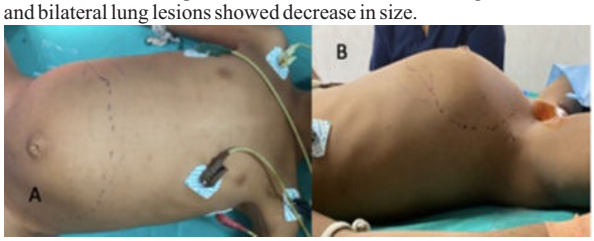

Figure 1: Clinical photograph of abdomen showing distension with huge mass visible occupying lower to central abdomen.

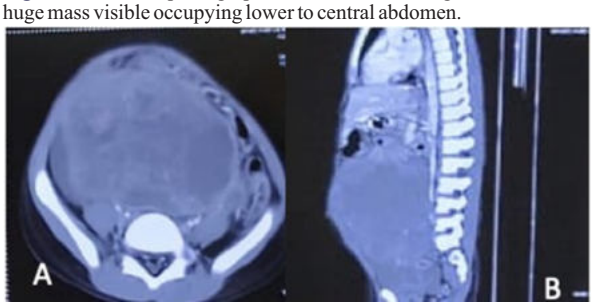

Figure 2: A, Axial and B, Sagittal section of CECT abdomen showing huge abdominopelvic mass

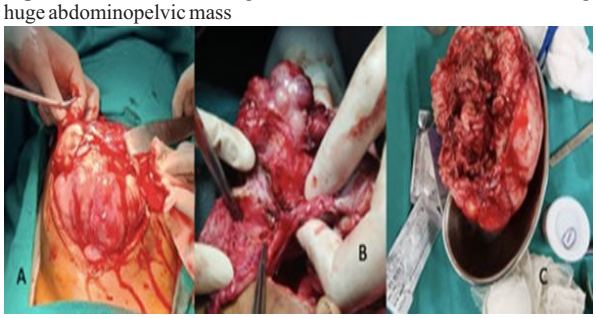

Figure 3: Intraoperative photographs A, huge tumor arising from bladder, B, Dense adhesions of the mass with postero-lateral wall of dome of bladder, C, Resected gross specimen showing rupture of tumour

DISCUSSION:

Rhabdomyosarcoma affecting the bladder/prostate constitutes 5% of the total RMS cases.[3] Rhabdomyosarcoma (RMS) stands as the predominant soft tissue sarcoma identified in children. In high-income countries, current treatment approaches yield an overall cure rate ranging from 70 to 80%, showcasing considerable variability across different risk groups. Conversely, low- and middle-income countries report less than a 50% overall survival, highlighting suboptimal outcomes in these regions.[4] The existing risk stratification system relies on data derived from the Intergroup Rhabdomyosarcoma Study (IRS) trials I-IV. Histology plays a crucial role in this stratification, and two histologic subtypes, embryonal RMS (ERMS) and alveolar RMS (ARMS), consistently serve as predictors of outcomes. Recent research has emphasized the significance of molecular characteristics, highlighting the critical prognostic role of FOXO1 fusion status, second only to metastatic status.[5]

Bladder and prostate is considered unfavourable site among genitourinary RMS. The primary objective of surgery for Rhabdomyosarcoma (RMS) is the complete removal of the tumor, while preserving cosmesis and function to the greatest extent possible. Achieving a complete removal without any microscopic disease offers the best chance of a cure. The surgical approach is determined by factors such as the primary site, tumor size, presence of lymph nodes, and distant metastases. For unresectable microscopic or gross residual disease, marking with titanium clips in the tumor bed is essential to guide re-excision and subsequent radiotherapy if needed, as was done in this case. Since most tumors are typically unresectable at presentation, chemotherapy with or without radiotherapy is often administered initially to reduce tumor size, facilitating a safe and complete surgery later on.[6] There is role of open incsional biopsy before commencing the chemotherapy. In our case, tumor was growing rapidly with imminent abdominal compartment syndrome affecting daily life of patient. So we planned for upfront resection followed by chemotherapy and radiotherapy.

The involvement of regional lymph nodes is a significant unfavorable prognostic factor. Radiation therapy (RT) is recommended to enhance local control in cases where patients have microscopic or gross residual tumor.[2] Brachytherapy has emerged as a viable alternative to external beam radiotherapy(EBRT) to minimise long term sequelae and toxicity to genitourinary and gastrointestinal system.[7]

In our case, we planned for an upfront surgery as the child had presented with a rapidly growing mass over 2 weeks. On imaging we found that the mass was locally invasive as well as metastatic (to the lungs). Intra op there was tumour spillage due rupture of capsule of the tumour, and hence we have also marked the area with titanium clips. Post-surgery, after reducing the tumour burden, we have given chemotherapy (VAC regimen) and also planning to go for radiotherapy at the operative site.

CONCLUSION:

Rapidly progressing pediatric abdominal lumps can cause abdominal compartmental syndrome and therefore an expedited treatment plan which includes upfront surgery can be lifesaving.

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