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Paediatric Oncological Surprises - A Case Series – An Institutional Experience

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Abstract:
Aims*: To study the uncommon presentations of common tumours at uncommon sites in the department of paediatric surgery, Coimbatore medical college hospital*
Materials*: Prospective study for one year from 2016 to 2017.The mode of presentation, clinical features, treatment and follow up were noted in these cases.*
Results*: There were three cases in this series. One 3year old female child presented with florid grape like lesions in the vagina and urinary retention owing to the tumour pressure on the bladder had elevated Serum AFP level, MRI revealed extensive pelvic sidewall infiltrate and histopathology revealed an extragonadal yolk sac tumour of vagina. Child improved with chemotherapy. Another child aged 5years had a posterior bladder wall mass which threw a surprise of an extrarenal Wilms tumour. Neoadjuvant chemotherapy was given and further surgical exploration with post op chemotherapy and radiotherapy had complete resolution. Another 4-year child with a gluteal mass diagnosed to have Para chordoma with recurrence revealing the same pathology on excision. However, IHC was suggestive of a sclerosing Rhabdomyosarcoma with chemotherapy child improved well.*
Conclusions*: Common tumours can present at uncommon sites as malignancies can throw you surprise at any moment. Histological surprises are case bound and the importance of AFP in vaginal masses has to be emphasized. A combined team approach with a dedicated paediatric surgeon, a watchful pathologist and an oncologist can work wonders in paediatric malignancies.*
Keywords: *Yolk sac tumour, Wilms tumour, Rhabdomyosarcoma*

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I. Introduction

Cancer in children is uncommon; it represents only about 2% of all cancer cases. Leukaemia (26%) is the most common form of cancer in children, and brain tumour's (21%) are the most common solid tumour of childhood. Lymphomas (15%) are the next most common malignancy in children, followed by neuroblastoma (7%), Wilms' tumour (5%), soft tissue sarcomas (3%), germ cell tumour's (8%), osteosarcoma (6%), and retinoblastoma (2%).

Tumour have a particular most common site of occurrence. It throws surprises when it is diagnosed in uncommon sites

II. Material And Methods

Prospective study for one year from 2016 to 2017.The mode of presentation, clinical features, investigations, treatment and follow up were noted in these cases.

Case 1: A 1 year female baby presented with history of bleeding per vagina of one month duration. Per abdomen examination showed distended bladder (retention of urine), perineum examination showed - seropurulent discharge from vagina and per rectal examination showed mass anterior to rectum with distended vagina. As the bladder was palpable, an indwelling
catheter was placed. MRI done showed a Ill- defined irregular infiltrative mass - 24x27x39mm - upper 2/3 of vagina, cervix & lower uterine segment - infiltration to pelvic sidewall & pelvic floor (Figure.1)

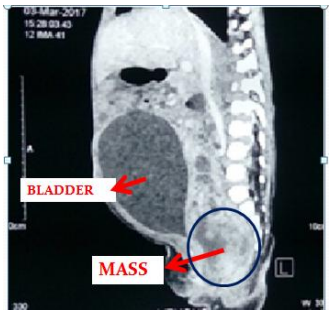

Figure.1

Child was evaluated with serum AFP showed value 20,000IU/ml, Cystoscopy done was normal, Genitoscopy showed a irregular friable growth arising from vagina

Biopsy from the growth was taken. Histopathology proved to be an Extragonadal yolk sac tumour / Endodermal sinus tumour (Schiller-Duval bodies were identified-Figure.2)

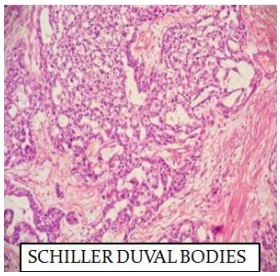

Figure.2

So an Extra gonadal Non sacral Yolk sac tumour (Vagina) -Stage III was diagnosed.Child was managed with Cisplatin,Etoposide & Bleomycin (PEB) regimen for six cycles once in 21days. On followup vaginal growth showed marked regression.

Case 2: A 4years malechild admitted with history of swelling in right thigh following a trivial injury one week back. Clinical examination showed a abnormally dilated veins over rightside of the aabdomen, with a diffuse swelling in right thigh(Figure.3).Perrectal examination showed a bulge at the anterior rectalwall.

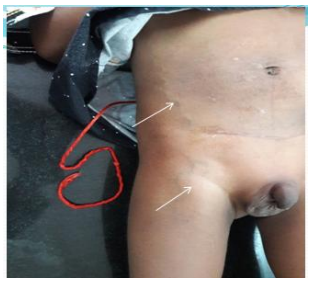

Figure.3

Ultrasound abdomen & pelvis showed a 5 x 5.5 cm growth posterior to bladder with thrombosis of IVC extending to both internal iliac veins and right external iliac vein ?Rhabdomyosarcoma. CT scan done showed a bladder mass with infitration to pelvis with IVC thrombus(Figure.4).

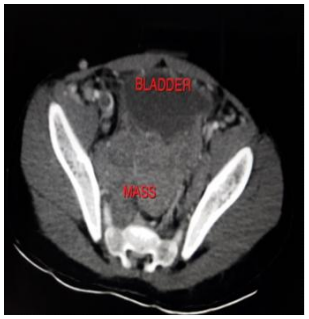

Figure.4

Cystoscopy done showed a extrinsic bladder compression.Laparotomy done showed a diffuse mass 5x4.5cm posterior to bladder compressing the rectum.Biopsy from the mass with sigmoid loop colostomy was done.Histopathology proved to be a Extrarenal Wilms tumour. So the child was managed with Chemotherapy(NWTS regimen),after 10^{th} week relapaotomy and excision of residual tumour was done. Post operative radiotherapy - followed by completion chemotherapy and colostomy closure done.

Case 3:A 3years male child admitted with a swelling in the left gluteal region of 1year duration.On examination a firm intramuscular,nontender,circumscribed swelling 4 x 4 cm over left gluteal region. Excision biopsy done(Specimen -figure.5) and histopathology reported as Benign Myoepithelioma (parachordoma).
On short period folowup child developed recurrence at the samesite.Ultrasound done showed a 6 x 4.5 cm mixed echogenic mass - muscle and subcutaneous plane ? Soft tissue tumour .So widelocal excision was done .

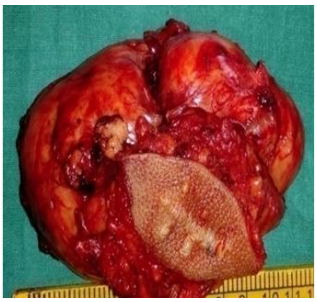

Figure.5

Histopathology reported as Malignant myoepithelioma / mixed tumour of soft tissue,Immunohistochemistry done showed a Sclerosing Rhabdomyosarcoma(Myo D1 & Vimentin positive).Child managed with chemotherapy(Vincristine,Actinomycin-D and Cyclophosphamide alternting with Ifosfamide,Etoposide) 16cycles for 48weeks.

III. Discussion

Case 1: Paediatric malignant germ-cell tumour's (GCT) are rare tumours of childhood accounting for less than 3% of paediatric malignancies [1]. In children less than 3 years of age, the most common sites for GCTs are extragonadal and gonads [2]. Endodermal sinus tumour (EST) is the most common histologic subtype of GCT [3]. Endodermal sinus tumour (EST) of the vagina, an extremely rare paediatric malignancy primarily affecting infants [4, 5]. In this situation it has to be differentiated from a sarcoma botyroides [6], which is more common.
In the treatment of vaginal EST, primary conservative surgery in the form of partial vaginectomy and adjuvant chemotherapy are highly attractive methods of preserving reproductive and sexual function.
The serum AFP level is a useful marker for diagnosis and monitoring the recurrence of vaginal EST in infants.
Case 2: Wilms tumour is one of the most common solid neoplasia of the childhood and the most common tumour of Genito-urinary tract [7]. Extra renal sites for origin of Wilms's tumour are extremely rare.
The diagnosis of extra renal Wilms tumour is made only after a primary tumour of the kidneys has been ruled out with a secondary extra renal metastasis [8]. There are two types of extra renal Wilms's tumour [9] One arises in the line from the renal bed to the scrotum, supposedly from residual embryonic renal tissue [10]. The second consists of teratoma with nephroblastic tissue [11]. The exact embryological origin is not clear. Presently there is no staging for extra-renal Wilms's tumour.
The prognosis and clinical course of extra renal Wilms's tumour is parallel to those of intra renal Wilms's tumour [12] Extra renal Wilms tumour should be treated according to NWTS protocol.
The unique features in our patient were a growth posterior to bladder with thrombosis of IVC extending to both internal iliac vein and right external iliac vein
Case 3: Rhabdomyosarcoma is the most common soft tissue malignancy in children under the age of 15years and most frequently occurs in the head and neck.
There are three types of rhabdomyosarcomas recognized by- the Intergroup Rhabdomyosarcoma Study group: embryonal rhabdomyosarcoma (ERMS), alveolar rhabdomyosarcoma (ARMS) and pleomorphic rhabdomyosarcoma (PRMS) [13]. Botryoid and spindle cell rhabdomyosarcoma
are distinct subtypes of ERMS.
Sclerosing rhabdomyosarcoma (SRMS), has been found only in adults and may have a predilection for sites in the head and neck. Folpe et al [14] have argued that SRMS may be a new type or subtype of rhabdomyosarcoma.
Although not yet borne out in the literature, this may have unique treatment and
prognosis implications for the patient.

IV. Conclusion

Common tumours can present at uncommon sites can throw you surprise at any moment. Histological surprises are case bound and the importance of AFP in vaginal masses, IHC in rhabdomyosarcoma has to be emphasized.

A combined team approach with a dedicated paediatric surgeon, a watchful pathologist and an oncologist can work wonders in paediatric malignancies.

References

[1]. Rescoria FJ (1997) Germ cell tumors. Semin Pediatri Surg 6: 29–37
[2]. Einhorn LH, Donahue J (1977) Cis-diaminedichloroplatinum, vinblastine, and bleomycin combination chemotherapy in disseminated testicular cancer. Ann Int Med 87: 293–298
[3]. Davidoff AM, Hebra A, Bunin N, Shochat SJ, Schnaufer L (1996) Endodermal sinus tumor in children. J Pediatr Sur31: 1075–1078
[4]. Horng YC, Tsai WY, Lin KH (1994) Infantile endodermal sinus tumor presenting with vaginal bleeding: report of a case. J Formos Med Assoc 93: 164–166
[5]. Hwang EH, Han SJ, Lee MK (1996) Clinical experience with conservative surgery for vaginal endodermal sinus tumor. J Pediatr Surg 31: 219–222
[6]. Copeland IJ, Sneige N, Ordonez NG (1985) Endodermal sinus tumor of the vagina and cervix. Cancer 55: 2558–2565
[7]. Broeker BH, Calamone AA. Primary extrarenal Wilm's tumour in children. J Paediatr Surg 1989; 12:24.
[8]. Andrews PE, Kelalid PP. Extra renal Wilm's tumour; result of national Wilm's tumour study. J Paediatr Surg 1992;9;27.
[9]. Mang LL, Jagadessan K. Extra renal Wilm's tumour. Med J Malaysia1983; 28:134-36.
[10]. Bhajekar AB, Joseph M, Bhat HS. Unattached nephroblastoma, Br. J. Urol 1964;36:187
[11]. Malik TA, Malik GB, Diesh G. Retroperitoneal teratoma with nephroblastoma tissue as the main component. Int Surg 1967; 47:246-9.
[12]. Coppes MJ, Wilson PCG, Wenaeitzman S. Extra renal Wilm's tumour staging treatment and prognosis. J Clin Oncol 1991; 9:167-74.
[13]. Crist WM, Anderson JR, Meza JL, et al. Intergroup Rhabdomyosarcoma Study, IV: results for patients with nonmetastaticdisease. J Clin Oncol 2001; 19:3091–3102
[14]. Folpe AL, McKenney JK, Bridge JA, Weiss SW. Sclerosing rhabdomyosarcoma in adults. Am J Surg Pathol 2002; 26:1175–1183.

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