Extra-renal Wilms' Tumor in a child: A Case Report

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Abstract

Wilms' tumor is one of the commonest abdominal tumors of childhood, primarily a malignant renal tumor. However, extrarenal Wilms' tumor (ERWT) is a rare disease, and only a few cases have been reported in the pediatric age group. Here we report a child with retroperitoneal mass, who had ERWT.

Keywords

- Wilms' tumor
- Pediatric
- extrarenal

Introduction

Renal tumors account for 6% of all the malignancies in children and Wilma's tumor is the commonest. Extra-renal Wilms' tumor (ERWT) or Extra-renal Nephroblastomatosis is a rare condition and accounts for 0.5-1% of all Wilms' tumor cases. ERWT has been reported in

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the retroperitoneum, scrotum, inguinal region, and pelvis.[1-2] Due to rarity of ERWT, it's prognostic markers and specific pre-operative diagnostic tools can't be determined. To date, less than 100 cases of ERWT in pediatric age groups have been reported.[4] Mostly it presents as a mass and general symptom related to the mass. Here we present a case of ERWT in a male child having a retroperitoneal mass.

Case Report

A twoyears old male child presented with an abdominal mass on the left side for two months. This mass was gradually increasing in size with no associated pain. However, there was an associated history of weight loss and anorexia. A 12 x 8 cm mass was palpable on abdominal examination, which was occupying the left hypochondrium and left lumbar region. It was a non-tender, firm, smooth, and mobile mass. Its upper limit was not reachable; however lower limit was approachable. His baseline investigations were obtained, which were within normal ranges. Ultrasound revealed it arising from left kidney and havingboth solid and cystic components within it, however, right kidney was not visualized. CT scan of the abdomen showed this mass attached to the retroperitoneum, closely abutting the left kidney's upper pole and having foci of calcifications. There was hydronephrosis on the left side due to mass compression, and the right kidney was absent. His renal Diethylenetriamine pentaacetate (DTPA) scan revealed absent/non-functioning right kidney and reduced left kidney function. His serum Alfa fetoprotein level was in normal range. The patient was optimized and explored. Per-operatively, mass was lying close to the left kidney and had both solid and cystic components. There was a hydroureter, and both the left kidney and ureter were pushed by mass. Mass was completely separated from surrounding structures and was excised in toto **(figure 1A & figure 1B)**. His postoperative recovery remained uneventful. A biopsy report of the mass showed it to be ERWT **(figure 2)**. The patient is in our two years follow-up and doing well to date.

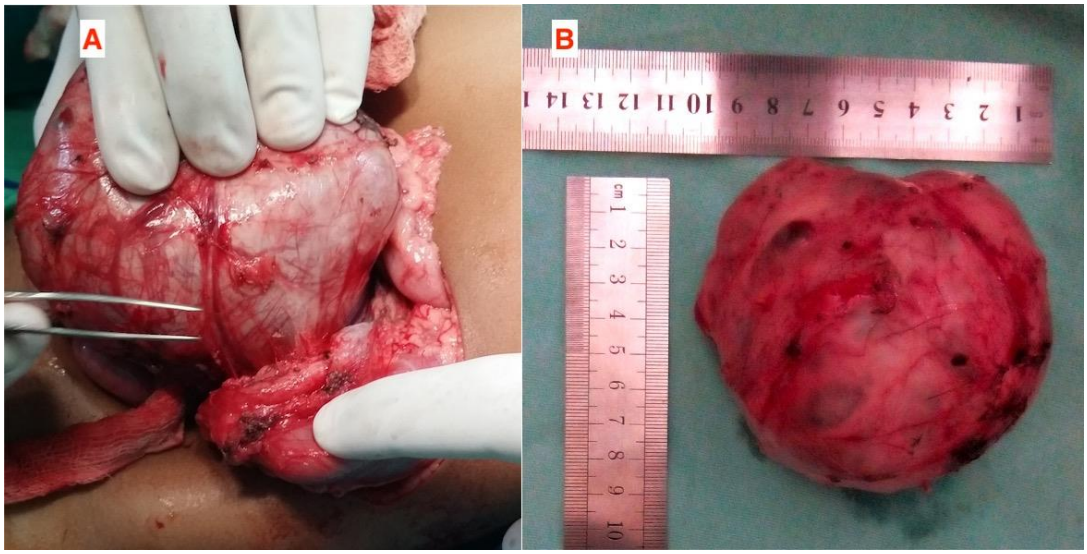

Figure 1:
A) Intra-operative picture of the mass
B) Mass excised in toto

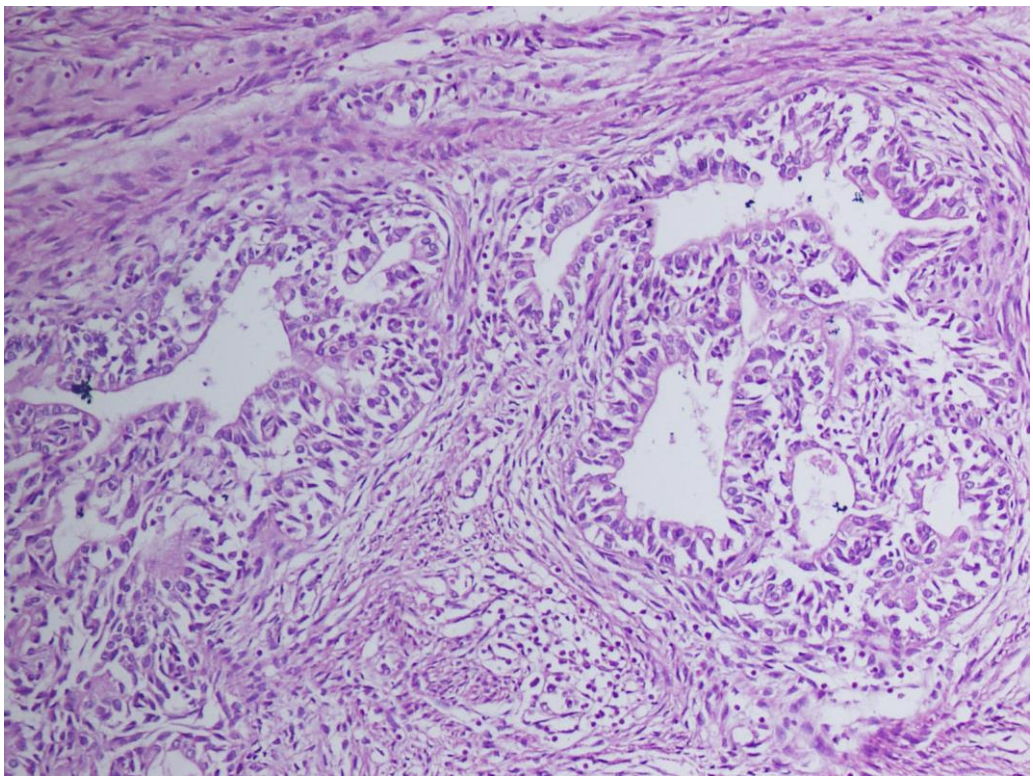

Figure 2: Histopathological picture of the mass

Discussion

Wilms' tumor is one of the most common abdominal malignancies of childhood; ERWT, on the other hand, is a rarity. The first case of ERWT was reported by Moyson in 1961.[1] Most cases that have been reported in literature, are predominately present in the retroperitoneum. Other reported sites for ERWT include sacrococcygeal region, paravertebral region, inguinal canal, ovary, uterus, paratesticular region, mediastinum and urinary bladder.[3-4]

Although this tumor's exact origin is still hazy, the most plausible hypothesis includes that of origin from ectopic metanephric blastema.[5] These ectopic cells may reside at any site and lead to ERWT. There is also a theory of romance of persistent embryoic potential cells (Connheims' cell rest theory). There are other situations in which ERWT may be found like as a metastatic disease or a nephroblastoma originating in a teratoma.[1]

ERWT are more common in females, with a female:male ratio of 56:44 and occurs within the first decade of life. Patients with ERWT present usually in range of 2 months to 10 years; contrary to the age of Wilms' tumor being 44 months.[1-6] The oldest case reported in literature of ERWT was in a 77 years old female who had uterine ERWT.[1]

ERWT usually presents as an asymptomatic abdominal mass with insidious growth. There is no specific presenting feature to it and usually they depend upon the site of origin.[5-7-6] In the indexed case, child had painless abdominal mass, causing hydronephrosis due to compression of the ureter and kidney by mass.

Investigating this tumor may reveal elevated tumor markers like alpha-fetoprotein and beta-HCG. As for imaging modalities, the initial and the most commonly used radiological intervention is ultrasonography. Sonograms usually demonstrate well defined, smooth mass of renal origin with uniform echogenicity. Contrast-enhanced CT scans provide a better and clearer picture of the tumor's quality and extension, highlighting the usual picture of a heterogeneous infiltrative mass with vessel encasement and calcification. Bony metastasis is common, for which bone scans can be useful.[2-9-11]

The differential diagnosis for ERWT (of cystic variety) includes cystic extragonadal germ cell tumors, neurogenic tumors, and, rarely, myxoid liposarcoma.[2-10]

Unfortunately, being an aggressive malignant tumor, it carries the potential of distant metastasis, and local recurrences. The cystic variety of this tumor has a better prognosis than solid ones.[3-4]

Ultimately, renal and extra-renal Wilms tumors can only be confirmed on histopathological evaluation, as in our case. The diagnosis of ERWT is usually difficult. As per National Wilms' Tumor Study Groups, three components are necessary inorder to make a diagnosis of ERWT. These include: 1) tumor should not be present in both kidney, 2) there must be a triphasic pattern in histopathology including undifferentiated blastemal, epithelial and mesenchymal components, and 3) no teratoma or renal cell carcinoma detected on histopathology.[1-4]

Approximately 200 cases of ERWT in both adults and children have been reported.[1] Shojaeian et al. reported 80 childhood ERWT cases from 1961-2015.[4] We did a literature review from 2015 onwards and retrieved 18 more cases. We also found four case[12-14] in the literature which were reported before 2015, but were missed by Shojaeian. Their summary has been given in **Table 1**.

Table 1:

Sr. No.	Author	Year	Age	Gender	Location
1.	Lakhkar et al [13]	2004	2 years	Male	Right iliac fossa
2.	Dabus et al [14]	2004	2 years	Female	Retroperitoneal right supra-renal
3.	C Ramachandra et al [12]	2007	4 years	Male	Retroperitoneal right lumbar
4.	C Ramachandra et al [12]	2007	3 years	Female	Retroperitoneal right supra-renal
5.	Thakkar et al [15]	2015	5 years	Female	Right ovary
6.	Park et al [16]	2016	4 years	Female	Inguinal canal
7.	Igbaseimokumo U et al [17]	2017	Newborn	Female	Lumbar Spine
8.	Ozkisacik et al [8]	2017	3 years	Male	Retro-vesical
9.	Wabada S et al [5]	2017	2 years	Male	Retroperitoneal mass
10.	Itoshima R et al [3]	2017	4 years	Male	Retroperitoneal

11.	Chin Tan et al [7]	2018	2 years	Female	Pelvic mass with skeletal and glial differentiation
12.	Tang H et al [18]	2018	2 years	Male	Retroperitoneal mass
13.	Tang H et al[18]	2018	2 years	Female	Mesentry
14.	Tan GC et al [7]	2018	2 years	Female	Pelvis
15.	Groth et al [19]	2019	2 years	Male	Para-testicular mass
16.	Sindhu et al [1]	2019	6 years	Male	Urinary bladder
17.	Liang H et al [6]	2020	5 years	Male	Retroperitoneal mass
18.	Liang H et al [6]	2020	3.4 years	Female	Presacral
19.	Liang H et al [6]	2020	3.4 years	Female	Duplicate sigmoid colon
20.	Liang H et al [6]	2020	9.8 years	Male	Retroperitoneal mass
21.	Liang H et al [6]	2020	2.8 years	Male	Gubernaculum Testis
22.	Filizoglu N et al [20]	2021	3 years	Female	Lumbar mass

Conclusion

We conclude that ERWT is a rare pathology, but it must be considered as one of the differentials among the retroperitoneal tumors. The diagnosis can only be confirmed with histopathology.

Ethical Considerations

This study is approved by the Institutional Ethics Committee of Yenepoya (Deemed to be University) with the Protocol No: YEC2/833.

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Conflict of interests

There are no conflicts of interest.

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