Chapter 6

Extrarenal Wilms' Tumor: Challenges in Diagnosis, Embryology, Treatment and Prognosis

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Abstract

Wilms' tumor is one of the most common childhood solid malignancies, which classically arises from primitive metanephric cells, but exceptionally it may arise in places other than kidneys. Extrarenal Wilms' tumor is a rare but challenging entity, considering its diagnosis,

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histopathology, staging, treatment, and prognosis. Diagnosis of extrarenal Wilms' tumor is always postsurgical, which may jeopardize treatment planning and consulting with parents in the first step. The histopathology of Wilms' tumor is very confusing. While most authors believe that it arises from primitive ectopic nephrogenic rests, teratoid Wilms' tumor leads to the debate whether this tumor is neoplastic or embryonic. Staging of extrarenal Wilms' tumor is also a challenge when we consider the National Wilms' Tumor Study (NWTS) recommendations; all these tumors should be considered as stage II or higher as they are beyond the renal capsule. This will mandate chemotherapy for all patients while most of the reported cases have a favorable histology, and long-term tumor-free survival has been reported even with exclusive surgery in some case reports. Although treatment strategies for extrarenal Wilms' tumor are the same as those for renal Wilms' tumor, different locations and neighboring organs may invoke special considerations and scenarios while planning for surgery and adjuvant therapies. Consulting with the parents is also a problem, considering the rarity of the disease and limited publications. In this chapter, we discuss all these topics in detail after a systematic review of extrarenal Wilms' tumor cases to date in order to provide a clear perspective for confronting this rare disease.

Key words: Extrarenal; Nephroblastoma; Pediatrics; Wilms' tumor

Introduction

Nephroblastoma or Wilms' tumor is one of the most common childhood malignancies, which accounts for almost 95% of renal malignancies in pediatrics. Extrarenal nephroblastoma is a rare entity, which was first described by Moyson et al. (1) in 1961. The estimated rate of occurrence of nephroblastoma outside the kidneys is almost 0.5 to 1% of all cases of Wilms' tumor. Extrarenal Wilms' tumor (ERWT) occurs mostly in childhood; however, it is also rarely reported in adults (2). Apart from the primary ERWT, nephroblastoma may be observed outside the kidneys in two other situations: metastatic disease and nephroblastoma arising in a teratoma; therefore, in the case of ERWT, it is mandatory to evaluate the kidneys for primary tumor preoperatively and search the whole specimen for any teratoid element postoperatively (3).

We reviewed all the reported childhood ERWTs (under 14 years), excluding those arising from teratomas (teratoid Wilms' tumor). The results are summarized in Table 1. Among 80 reported ERWT cases, more than 60% were younger than 4 years and a female predominance was observed while the female-to-male ratio was 3:2 (4).

The association of ERWT with a horseshoe kidney has been reported previously, and almost 7% of the reported ERWTs were found to be associated with the horseshoe kidney. Dysraphism is the second commonly found abnormality among ERWT patients (5).

Table 1. Review of the reported childhood ERWTs in literature

Author	Year	Gender	Age	ERWT location	Stage	Treatment	Follow-up
1. Moyson et al. (1)	1961	F	3	Mediastinal	II	Surgery + chemo	NA
2. Bhajkar et al. (6)	1964	M	2	Retroperitoneal	II	Surgery + chemo	NA
3. Edelstein et al. (7)	1965	M	3	Retroperitoneal	II	Surgery + chemo + RAD	2
4. Wu and Garcia (8)	1971	F	7	Pelvic	IV	Surgery + chemo + RAD	1
5. Thompson et al. (9)	1973	F	4	Inguinal	III	Surgery + chemo + RAD	2
6. Thompson et al. (9)	1973	M	3	Inguinal	IV	Surgery + chemo + RAD	0.5
7. Akhtar et al. (10)	1977	M	0.2	Inguinal	II	Surgery + chemo	1.5
8. Gaikwad et al. (11)	1977	M	0.2	Retroperitoneal	II	Surgery	0.5
9. Madanat et al. (12)	1978	F	9	Mediastinal	II	Surgery + chemo + RAD	3
10. Madanat et al. (12)	1978	M	0.3	Inguinal	NA	Surgery + chemo	2
11. McCauley et al. (13)	1979	F	4	Retroperitoneal	NA	Surgery + chemo + RAD	4
12. Aterman et al. (14)	1979	F	5	Retroperitoneal	NA	Surgery + chemo + RAD	0.7
13. Orlowski et al. (15)	1980	M	3.5	Paratesticular	II	Surgery	11
14. Fried et al. (16)	1980	M	3	Retroperitoneal	NA	Surgery + chemo	NA
15. Fernandes et al. (17)	1980	M	6	Retroperitoneal	III	Surgery + chemo + RAD	6
16. Johnson et al. (18)	1980	F	1	Retroperitoneal	NA	Surgery + chemo	1
17. Taylor et al. (19)	1980	M	0.5	Inguinal	NA	Surgery + chemo + RAD	0.5
18. Ho et al. (20)	1981	M	1.2	Paratesticular	I	Surgery	1
19. Bittencourt et al. (21)	1981	F	14	Female genital organs	III	Surgery + chemo + RAD	5.5

(*Continued*)

Table 1. Continued

Author	Year	Gender	Age	ERWT location	Stage	Treatment	Follow-up
20. Tamaro et al. (22)	1982	F	4	Retroperitoneal	NA	Surgery + chemo + RAD	1
21. Fernandes et al. (17)	1982	F	2	Retroperitoneal	II	Surgery + chemo	5
22. Adam et al. (23)	1983	M	10	Retroperitoneal	NA	NA	NA
23. Meng and Jagadeesan (24)	1983	M	3	Retroperitoneal	I	Surgery	1
24. Lüchtrath et al. (25)	1984	F	1.2	Inguinal	NA	Surgery + chemo	1.3
25. Bell et al. (26)	1985	F	13	Female genital organs	I	Surgery	9.5
26. Naito et al. (27)	1985	F	3	Retroperitoneal	NA	Surgery + chemo	2.3
27. Fernandes et al. (17)	1988	F	2	Retroperitoneal	II	Surgery + chemo	1
28. Lai et al. (28)	1888	F	3	Inguinal	II	Surgery + chemo	1.5
29. Narasimharao et al. (29)	1989	F	NA	Retroperitoneal	NA	NA	NA
30. Broecker et al. (30)	1989	F	0.9	Pelvic	II	Surgery + chemo	1
31. Fernandes et al. (17)	1989	M	6	Retroperitoneal	II	Surgery + chemo + RAD	7
32. Broecker et al. (30)	1989	F	2	Retroperitoneal	II	Surgery + chemo + RAD	7
33. Wakely et al. (31)	1989	F	4	Female genital organs	II	Surgery + chemo + RAD	6
34. Broecker et al. (30)	1989	F	2	Retroperitoneal	IV	Surgery + chemo	1.3
35. Wakely et al. (31)	1989	F	1.5	Retroperitoneal	II	Surgery + chemo + RAD	6
36. Strand et al. (32)	1990	M	12	Inguinal	III	Surgery + chemo	NA
37. Simha and Doctor (33)	1991	F	3	Inguinal	III	Surgery + chemo + RAD	NA

(*Continued*)

Table 1. Continued

Author	Year	Gender	Age	ERWT location	Stage	Treatment	Follow-up
38. Andrews et al. (34)	1992	F	NA	Lumbosacral	II	Surgery + chemo	1.4
39. Andrews et al. (34)	1992	M	NA	Retroperitoneal	II	Surgery + chemo	0.7
40. Andrews et al. (34)	1992	F	NA	Lumbosacral	II	Surgery + chemo	6.5
41. Andrews et al. (34)	1992	M	NA	Retroperitoneal	IV	Surgery + chemo + RAD	2
42. Andrews et al. (34)	1992	F	NA	Lumbosacral	III	Surgery + chemo + RAD	4
43. Andrews et al. (34)	1992	F	NA	Retroperitoneal	I	Surgery + chemo	3
44. Andrews et al. (34)	1992	M	NA	Retroperitoneal	II	Surgery + chemo	2
45. Andrews et al. (34)	1992	F	NA	Pelvic	II	Surgery + chemo	0.7
46. Suzuki et al. (35)	1993	M	2	Retroperitoneal	II	Surgery + chemo	NA
47. Rasheed et al. (36)	1993	M	3	Retroperitoneal	III	Surgery + chemo + RAD	7
48. Rasheed et al. (36)	1993	F	4	Retroperitoneal	III	Surgery + chemo + RAD	2
49. Fahner et al. (37)	1995	F	2.5	Lumbosacral	II	Surgery + chemo	1
50. Arkovitz et al. (38)	1996	M	3.5	Inguinal	III	Surgery + chemo + RAD	NA
51. López Cubillana et al. (39)	1997	F	2	Retroperitoneal	II	Surgery + chemo	3
52. Kapur et al. (40)	1998	F		Retroperitoneal	NA	Surgery + chemo	3
53. Kapur et al. (40)	1998	F	NA	Retroperitoneal	NA	Surgery + chemo	0.7
54. Benatar et al. (41)	1998	F	11	Female genital organs	II	Surgery + chemo	NA
55. Massarelli et al. (42)	1999	F	2	Female genital organs	III	Surgery + RAD	2.5

(*Continued*)

Table 1. Continued

Author	Year	Gender	Age	ERWT location	Stage	Treatment	Follow-up
56. Iraniha et al. (43)	1999	F	12	Female genital organs	II	Surgery + chemo	1
57. Govender et al. (44)	2000	M	4	Lumbosacral	III	Surgery + RAD	NA
58. Babin et al. (45)	2000	F	13	Female genital organs	III	Surgery + chemo + RAD	5
59. Arda et al. (46)	2001	F	5	Lumbosacral	III	Surgery + chemo + RAD	NA
60. Oner et al. (47)	2002	F	3.5	Female genital organs	II	Surgery + chemo	7
61. Yunus et al. (48)	2003	NA	NA	Lumbosacral	NA	NA	NA
62. Cojean et al. (49)	2003	M	0.2	Retroperitoneal	III	Surgery + chemo	NA
63. Ngan et al. (50)	2009	F	6	Retroperitoneal	II	Surgery	1
64. Cooke et al. (51)	2009	M	1.2	Inguinal	II	Surgery	2
65. Jeong et al. (52)	2011	M	9	Inguinal	III	Surgery + chemo + RAD	NA
66. Teerthanath (53)	2011	F	6	Retroperitoneal	II	Surgery + chemo	4
67. Chowhan (54)	2012	M	1.3	Retroperitoneal	II	Surgery + chemo	NA
68. Armanda et al. (2)	2012	F	0.1	Lumbosacral	I	Surgery + chemo	2
69. Yamamoto et al. (55)	2012	M	NA	Paratesticular	NA	NA	NA
70. Li et al. (56)	2012	F	2	Pelvic	III	Surgery + chemo + RAD	3
71. Marwah et al. (57)	2012	F	1.2	Retroperitoneal	II	Surgery + chemo	NA
72. Hiradfar et al. (58)	2012	F	9	Inguinal	II	Surgery + chemo	3
73. Baskaran (59)	2013	M	3	Retroperitoneal	II	Surgery	NA
74. Rojas et al. (60)	2013	M	2	Lumbosacral	II	Surgery + chemo	NA

(*Continued*)

Table 1. Continued

Author	Year	Gender	Age	ERWT location	Stage	Treatment	Follow-up
75. Morandi et al. (61)	2013	M	3	Paratesticular	I	Surgery	2
76. Goel et al. (62)	2014	NA	NA	Retroperitoneal	NA	Surgery	NA
77. Wu et al. (63)	2014	M	0.8	Retroperitoneal	II	Surgery	0.5
78. Wu et al. (63)	2014	M	1.5	Inguinal	II	Surgery	0.5
79. Al-Nsoor et al. (64)	2014	F	1.7	Retroperitoneal	II	Surgery	NA
80. Kumar et al. (5)	2015	F	7	Retroperitoneal	NA	Surgery	0.8

Age and follow-up time are in years.
NA, data not available; RAD, radiotherapy.

Embryogenesis

Embryonic nephrogenic tissue normally differentiates into metanephric blastema, which is considered as the precursor of nephrons and mesenchymal stroma. The pronephros is developed during the third gestational week in the cervical region, extending to the more caudal parts, the cloaca. The upper part of pronephros regresses, and in the caudal part, it gives rise to the mesonephric duct that persists in two lateral foci. While the mesonephric duct extends to the cloaca, it enters to the metanephric blastema that surrounds the ureteric bud to develop the kidneys.Therefore, the progenitors of renal tissue goes through a journey from the cranial to the caudal part of the embryo, and during the longitudinal growth of the fetus, the intrapelvic kidneys ascend up to their expected level in the flank.

It is believed that the Wilms' tumor is the result of a developmental abnormality in the metanephric blastema. Persistent metanephric tissue after the 36th week of gestation could be the precursor of nephroblastoma. Nephrogenic rests may be observed anywhere in the craniocaudal migration line of primitive mesonephros and metanephros cells (Figure 1).

The pathogenesis of ERWT remains elusive, and several theories are discussed about it. Some authors have suggested that ERWT may arise from teratomas although later classifications divided teratoid Wilms' tumor and a true ERWT into two different entities while a true primary ERWT lacks any teratogenic element.

The most widely accepted hypothesis for the pathogenesis of ERWT suggests that the ectopic nephrogenic rest develops into a nephroblastoma. It is well known that the persistent

intrarenal fetal nephrogenic blastemal tissue may undergo oncogenic mutation and develop nephroblastoma. Several reports pointed to the observation of ectopic nephrogenic rests, especially in inguinal or retroperitoneal and lumbosacral regions (63).

The hypothesis of ectopic blastematous cells explains the development of ERWT in the craniocaudal migration pathway of primitive metanephros cells. The observation of WT1 gene in 25% of ERWT supports the oncogenic mutation of nephrogenic rests causing ERWT (65). The question is how we can explain the relatively high prevalence of ERWT in the inguinal region. Some genital structures such as Gartner's duct, seminal vesicles, vas deferens, and epididymis are differentiated mesonephric ducts that could explain the presence of ectopic nephrogenic rests as a precursor of ERWT in the inguinal region. Currently, several evidences support the hypothesis of ectopic nephrogenic blastemal cells causing the ERWT, which help us in better understanding of Wilms' tumor pathogenesis. Early diagnosis of nephrogenic rests outside the kidneys as a precancerous tissue will mandate close observation and prompt intervention while facing any evidence of atypia or malignancy (66).

Pathology

A classic microscopic feature of Wilms' tumor consists of the triphasic pattern that includes mesenchymal, epithelial, and blastemal elements. Histologic diagnosis of ERWT is supported by the observation of classic triphasic histology in the absence of any teratoid component. The presence of heterotopic teratomatous elements in more than 50% of total microscopic field suggests the diagnosis of teratoid Wilms' tumor, which is a quite different entity in the pathogenesis and embryology with germ cell origin. While confronting an extrarenal nephroblastoma, whole specimen should be examined in multiple cuts to exclude the diagnosis of teratoid Wilms' tumor that accounts for half of the reported extrarenal nephroblastic malignancies (67).

As discussed previously, ectopic nephrogenic rest is believed to be the precursor of Wilms' tumor, and histologic discrimination between these two is always a challenge (68). The presence of disordered structures, atypical mitosis, and marked pleomorphisms indicate the presence of Wilms' tumor in contrast with proliferative nephrogenic rests without atypia (63).

Considering the histologic findings, ERWTs could be classified as favorable or unfavorable. The review of reported cases shows predominant favorable histology among ERWTs. Beckwith and Palmer (69) proposed the criteria for ERWT pathological diagnosis that include the documented classic triphasic Wilms' pattern outside kidneys in the absence of teratoid or anaplastic elements while both kidneys are tumor free in imaging (Figure 2).

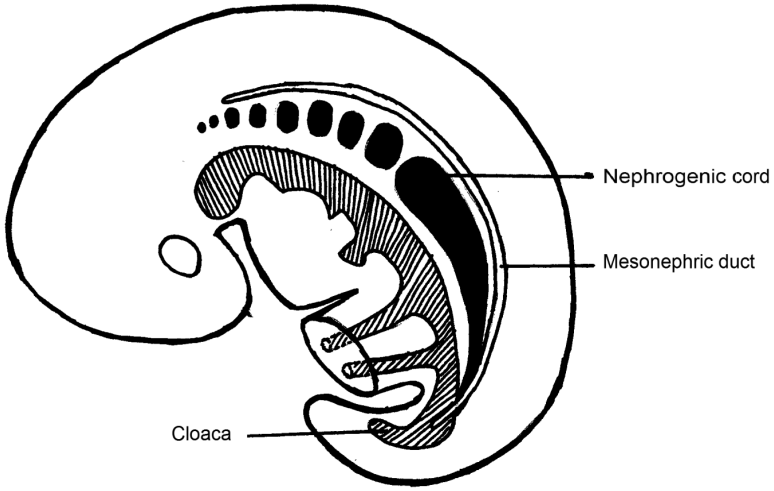

Figure 1. Schematic sagittal view of a 4-week-old embryo. Mesonephros consists of mesonephric duct and nephrogenic cord (Drawing by R. Shojaeian originally).

Clinical presentation and diagnosis

Clinical presentation of ERWTs depends on the location and stage of the tumor. As the symptoms are nonspecific and mostly due to the mass effect of tumor, diagnosis of ERWT often becomes apparent postoperatively. Common ERWT sites include retroperitoneum, inguinal area, lumbosacral and pelvic, female genital organs (uterus, cervix, vagina, and ovaries), mediastinum and chest wall, and spermatic cord and paratesticular region. Like classic Wilms' tumor, ERWT commonly manifests with asymptomatic mass or nonspecific symptoms, such as abdominal pain and discomfort, weight loss, and urologic or gynecologic symptoms. Symptoms related to the tumor site and size, such as inguinal mass that resembles hernia or lymphadenitis, vaginal bleeding or discharge, hematuria or dysuria, dyspnea, spinal cord compression signs, and even paraplegia (61, 3) may be observed.

Relation between ERWT and horseshoe kidney is suggested as we observed the coexistence of horseshoe kidney and ERWT in almost 13% of all previously reported cases. This may be explained by the higher chance of ectopic primitive renal tissue among patients with abnormal migration of nephrogenic cells, so the diagnosis of ERWT must be kept in mind while confronting with an abdominal mass in a patient with horseshoe kidney (59).

Ultrasound study is usually the first paraclinical step in the evaluation of an abdominal or pelvic mass. Further imaging such as computed tomography (CT) or magnetic resonance study may be needed in some cases. However, ERWT does not have a pathognomonic radiologic image, and the diagnosis of ERWT is almost always postoperative. After pathologic

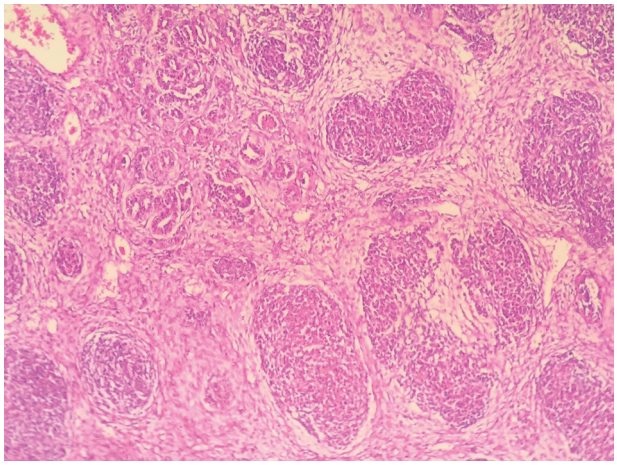

Figure 2. Microscopic evaluation of the specimen revealed composition of sheets, which were randomly arranged and tightly packed. Small blue cells were arranged in serpiginous aggregates (blastemal component), sharply circumscribed by focal spindling and intervening collagenous bundles apart from the surrounding stromal elements. There are also a few small tubules lined by primitive cuboidal cells and a small area of nephrogenic rest at the periphery. Pathological features suggested extrarenal Wilms' tumor (from Authors' personal archive).

confirmation of ERWT, both kidneys should be evaluated precisely with multislice spiral CT images to exclude any intrarenal tumor. CT scan with intravenous and oral contrast is often indicated and helpful to evaluate the tumor location and its resectability. Magnetic resonance imaging may also be helpful in paraspinal and thoracic tumors, especially with cord compression symptoms (64) (Figure 3). Recently, due to probable hazardous effects of CT scan especially in pediatric imaging, it is preferred to use MRI for evaluation of tumors in pediatric.

Tumor markers such as AFP (alpha-fetoprotein) and βHCG (beta-human chorionic gonadotropin) are useful to discriminate ERWT from other pediatric neoplasms such as the highly frequent germ cell tumors in childhood. Although a systemic search should be done for the tumor spread, ERWTs rarely metastasize. The most common sites of metastasis include lungs and liver. Three percent of the reported ERWT cases were metastatic based on our review.

Treatment

It is widely believed that National Wilms' Tumor Study (NWTS) system could be applied for ERWTs staging, while stage I definition should be modified as a localized tumor that could be completely excised with microscopic clear margins, no residue, and no tumor rupture during surgery; otherwise, there would not be any stage I ERWT as all tumors are located

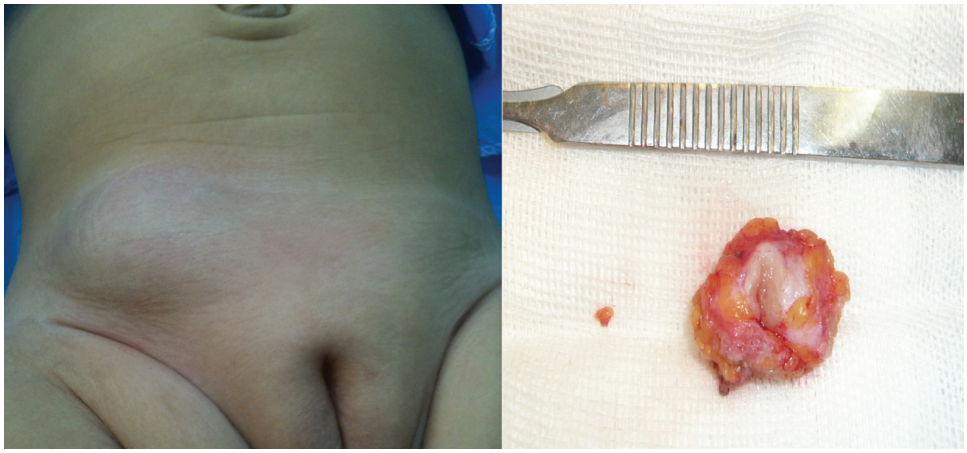

Figure 3. Left, Clinical manifestation of an inguinal ERWT. Right, Gross macroscopic view of inguinal ERWT (from Authors' personal archive).

beyond the borders of the kidneys. Although ERWT is uncommon, it should be kept in mind as a differential diagnosis of retroperitoneal or inguinal masses in childhood. Surgical approach depends on the location and surrounding structures. Rare cases of retroperitoneal or thoracic ERWT with intraspinal component and neurologic symptoms should be managed by multidisciplinary approach promptly to prevent irreversible neurologic sequels (64).

The role of intraoperative frozen section in an unidentified childhood mass or ERWTs has not been discussed clearly before and is not considered as a part of surgical principle, while total excision is the mainstay of treatment in most pediatric solid tumors when applicable (50).

Surgical excision remains the key step in the treatment of ERWT, especially when performed radically (70). Regional lymph node sampling is a part of the surgical principle as that for classic renal Wilms' tumor. Careful inspection of solid organs such as kidneys or liver and also peritoneum for tumor implants is recommended in abdominal ERWTs. Adjuvant chemotherapy is recommended for all ERWT cases postoperatively in spite of favorable histopathology in most of them. However, there are a few cases of successful treatment of stage I ERWTs with pure surgery.

Chemotherapy regimen is determined by histology and stage of the tumor, considering the NWTS protocols that consist of the administration of vincristine, actinomycin D, and doxorubicin. Regarding the current guidelines of NWTS, completely resected ERWTs with no evidence of tumor at or beyond the margins are considered as stage II and treated with vincristine and actinomycin D, while addition of doxorubicin will have benefits in stage III ERWTs.

Most ERWTs have favorable histology, but local recurrence is observed in about 11% of the reported cases, which is comparable with 15% predicted recurrence rate in classic renal Wilms' tumor with favorable histology. We found that 70% of ERWTs were in stage II and 23% in stage III, while distant metastasis was reported in 6% of patients. Two-year event-free survival of the reported ERWT cases was almost 85% and mortality rate was 5%, which are comparable with renal Wilms' tumor (60). Radiotherapy is reserved for unresectable tumors or for those with gross residue, recurrence, or metastasis (67). Bilateral ERWT has not been reported to date.

Conclusion

ERWT is considered a rare childhood malignancy with atypical presentations. The pathogenesis of ERWT becomes clearer by the popular theory, which suggests the heterotopic metanephric blastema as the precursor of ERWT while the diagnosis, staging, and treatment remain challenging. NWTS protocols are applied for ERWTs due to the rarity of the disease and lack of systematic data. We reviewed 87 reported childhood ERWT cases and observed favorable histology in most cases, which made the prognosis good and comparable to that of classic Wilms' tumor with the same stage and histology.

Conflict of Interests

The authors declare no potential conflicts of interest with respect to research, authorship and/or publication of this article.

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