An Astonishing Extrarenal Wilms Localisation; Spinal Cord

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Abstract

Wilms' tumor is one of the most common childhood solid malignancies, which accounts for almost 95% of renal malignancies in pediatrics, and classically arises from primitive metanephric cells. Exceptionally it may occur at places other than kidneys. The estimated rate of nephroblastoma outside the kidneys is almost 0.5 to 1% of Wilms' tumor cases. Extrarenal Wilms' tumor occurs mostly in childhood. In this article, we report a 3-year-old girl who first presented with spinal dysraphism and a mass in the lumbar spinal cord with a histopathological diagnosis of nephrogenic rest, and after one year, a Wilms tumor arose in this location. When extrarenal wilms tumor located in the spine is unlikely to be suspected preoperatively because it is embryologically less intuitive. Thus, we report this case of a congenital Wilms tumor associated with spinal dysraphism to increase awareness and describe this malignant tumor's clinical outcome.

Abstract: Wilms' tumor is one of the most common childhood solid malignancies, which accounts for almost 95% of renal malignancies in pediatrics, and classically arises from primitive metanephric cells. Exceptionally it may occur at places other than kidneys. The estimated rate of nephroblastoma outside the kidneys is almost 0.5 to 1% of Wilms' tumor cases. Extrarenal Wilms' tumor occurs mostly in childhood. In this article, we report a 3-year-old girl who first presented with spinal dysraphism and a mass in the lumbar spinal cord with a histopathological diagnosis of nephrogenic rest, and after one year, a Wilms tumor arose in this location. When extrarenal wilms tumor located in the spine is unlikely to be suspected preoperatively because it is embryologically less intuitive. Thus, we report this case of a congenital Wilms tumor associated with spinal dysraphism to increase awareness and describe this malignant tumor's clinical outcome.

Introductıon

Wilms' tumor is one of the most common childhood solid malignancies, which accounts for almost 95% of renal malignancies in pediatrics, and classically arises from primitive metanephric cells. Exceptionally it may occur at places other than kidneys. The occurrence of nephroblastoma outside the kidneys in the absence of a primary renal tumor is known as the extrarenal Wilms tumor (ERWT). ERWT is an uncommon entity that was first described by Moyson et al. in 1961(1). The estimated rate of nephroblastoma outside the kidneys is almost 0.5 to 1% of Wilms' tumor cases. Extrarenal Wilms' tumor occurs mostly in childhood; however, it is also rarely reported in adults (2) that typically involves the embryonic path of the developing kidneys and gonads. 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).

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 (4).

In this article, we report a 3-year-old girl who first presented with spinal dysraphism and a mass in the lumbar spinal cord with a histopathological diagnosis of nephrogenic rest, and after one year, a Wilms tumor arose in this location. ERWT is an unusual entity that typically involves the embryonic path of the developing kidneys and gonads. ERWT located in the spine is unlikely to be suspected preoperatively because it is embryologically less intuitive. Thus, we report this case of a congenital Wilms tumor associated with spinal dysraphism to increase awareness and describe this malignant tumor's clinical outcome.

Case Report

A 3-year-old girl was admitted to our clinic with difficulty in walking beginning 1.5 months before admission. We learned that she had been operated on five times since two months of age with dermal sinüs and tethered cord diagnoses. However, no pathological examination was done for the operation specimens.

Before 1.5 months, when she had difficulty in walking, she was admitted to our hospital, and an intradural mass (2.3x2x1.5 cm) was diagnosed in lumbosacral (L2-S2) spinal MRI (Fig 1). It was operated on, and the mass was totally excised in our neurosurgical department with a diagnosis of intradural abscesses, but the pathological diagnosis was a solid mass containing immature renal cells. Forty percent of it were blastemal nodules. The proliferation index was 2.8 %, and mitosis was rare (Fig 2).

After 1.5 months, the patient was admitted again with the complaints of the increase in difficulty in walking, standing, and balance.

The patient was hospitalized. He had difficulty in walking, and he was incontinent of urine and stool. Physical examination revealed weakness in the lower extremities (muscle strength was 1/5 bilaterally), and deep tendon reflexes were absent. The plantar response was bilateral flexor. Based on the clinical findings, spinal cord compression was considered, and spinal magnetic resonance imaging was performed. Lumbar magnetic resonance imaging showed a mass at the L2-S2 level involving the spinal cord (Fig 3). Urinalysis, renal, and liver function tests were normal. Abdominal ultrasonography and MRI were normal. Surgical resection of the tumor was performed, but the tumor mass could not be completely removed (Fig 4). The histopathological examination of the mass revealed triphasic histology Wilms' tumor in which blastemal, stromal, and epithelial elements were present (favorable histology) (Fig 5). According to the National Wilms' Study Group protocol, the tumor was accepted as Stage III.

The patient is six months follow after the surgery. Physical examination reveals the weakness in the right lower extremity (muscle strength 4/5). She has no incontinence. She is still being given chemotherapy and radiotherapy.

Discussion

ERWT in the spine is very rare. ERWT occurring within teratomas (5-10) is pathologically described as teratoid Wilms tumors and separate entities from the classic ERWT described in this case report. No elements of teratoma were found in our patient.

Posalaky et al. (11) described nephrogenic rests in the spine and suggested that these cells could undergo malignant transformation. There was no associated spinal dysraphism in their two reported cases, and the nephrogenic rests identified in the spine were benign with no evidence of ERWT. However, ten years later, Fahner et al. (12) described ERWT in the spine without spinal dysraphism.

The association of spinal dysraphism with ERWT is explained by the embryonic rest hypothesis of cancer development (13). This theory states that remnants of embryonic tissue occur in adults and children and that a change in the surrounding tissue would allow the embryonic tissue to resume proliferation and to produce masses of cells that resemble fetal tissues (14). Thus, it has been proposed that ERWT arises from pluripotent (mesenchymal) cell rests that undergo malignant transformation (13,14). Fernbach et al. (15), citing Grobstein's (16) study, suggested that the embryonic central nervous system is capable of inducing

nephrogenic differentiation in the embryonic mesenchyme from which the spine develops. Furthermore, Deshpande et al. (17) reported an ERWT within the dorsal lumbar spine's subcutaneous fat without associated spinal dysraphism, very similar in location to the nephrogenic rests described by Horenstein et al. (18), both of which had no associated spinal dysraphism.

The abortive formation of nephrogenic elements and the location of the tumor in the bifid spine outside the path of the developing kidneys, as in our case, supports the hypothesis that mesenchymal rests trapped in the wrong location and under aberrant stimulation develop into nephrogenic rests that undergo malignant transformation into ERWT. (13)

In a review of 34 cases of ERWT, Coppes et al. (19) analyzed therapy and survival. They suggested that all patients with ERWT receive postoperative chemotherapy, with the same regimen as for a renal Wilms tumor of the comparable stage. Sastri et al. (20) reviewed three additional cases and summarized a total of 48 cases of ERWT, and concurred with the recommendations of Coppes et al. (19). Based on these recommendations, our patient was treated according to the latest guidelines for renal Wilms tumor, following the National Wilms Tumor Study-V regimen for EE4A.

Conclusion

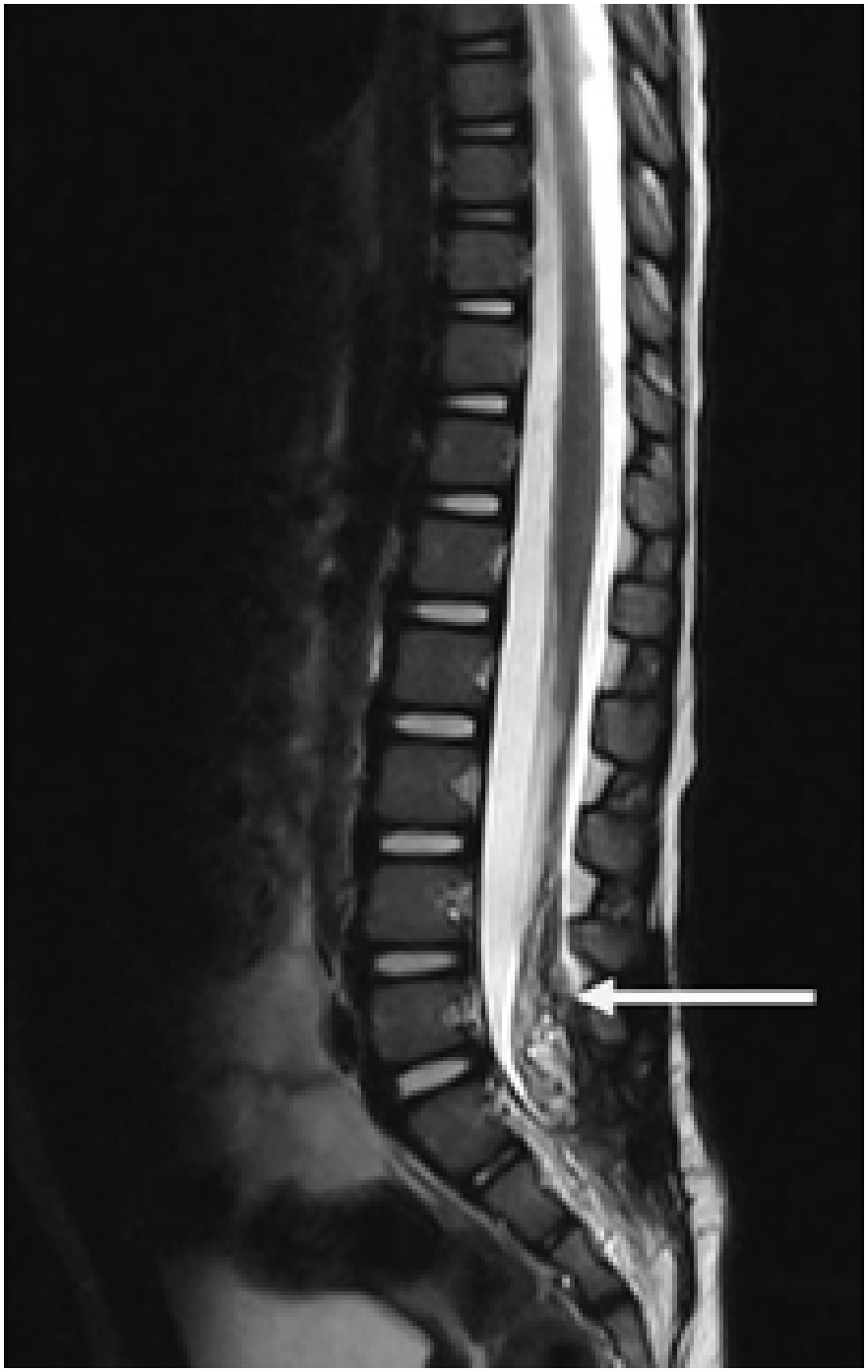
Here, we report an unusual case of ERWT. This report shed light on the debates of the pathogenesis of this tumor and that the embryonic rest theory explains the tumor's location away from the path of the embryonic kidney. Early surgical treatment of atypical masses in this location is recommended.

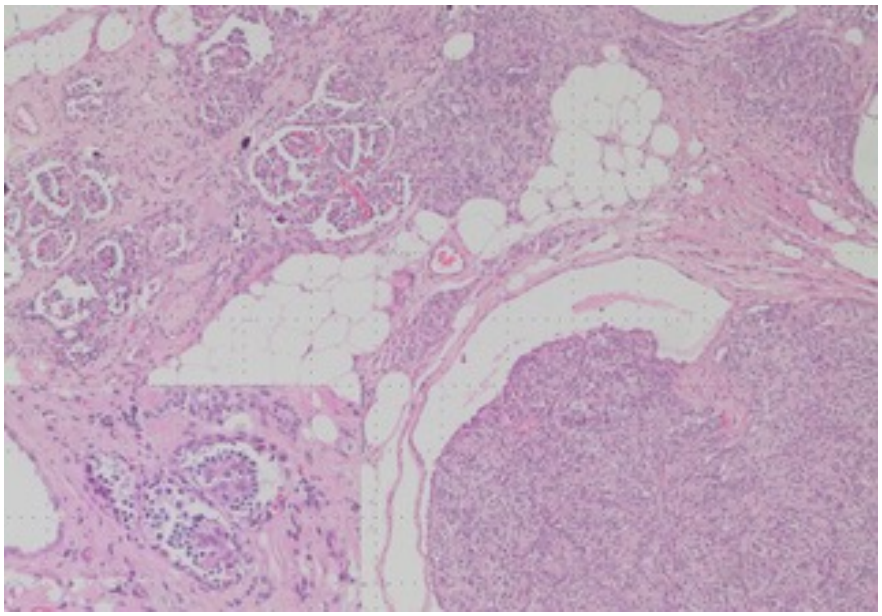
References

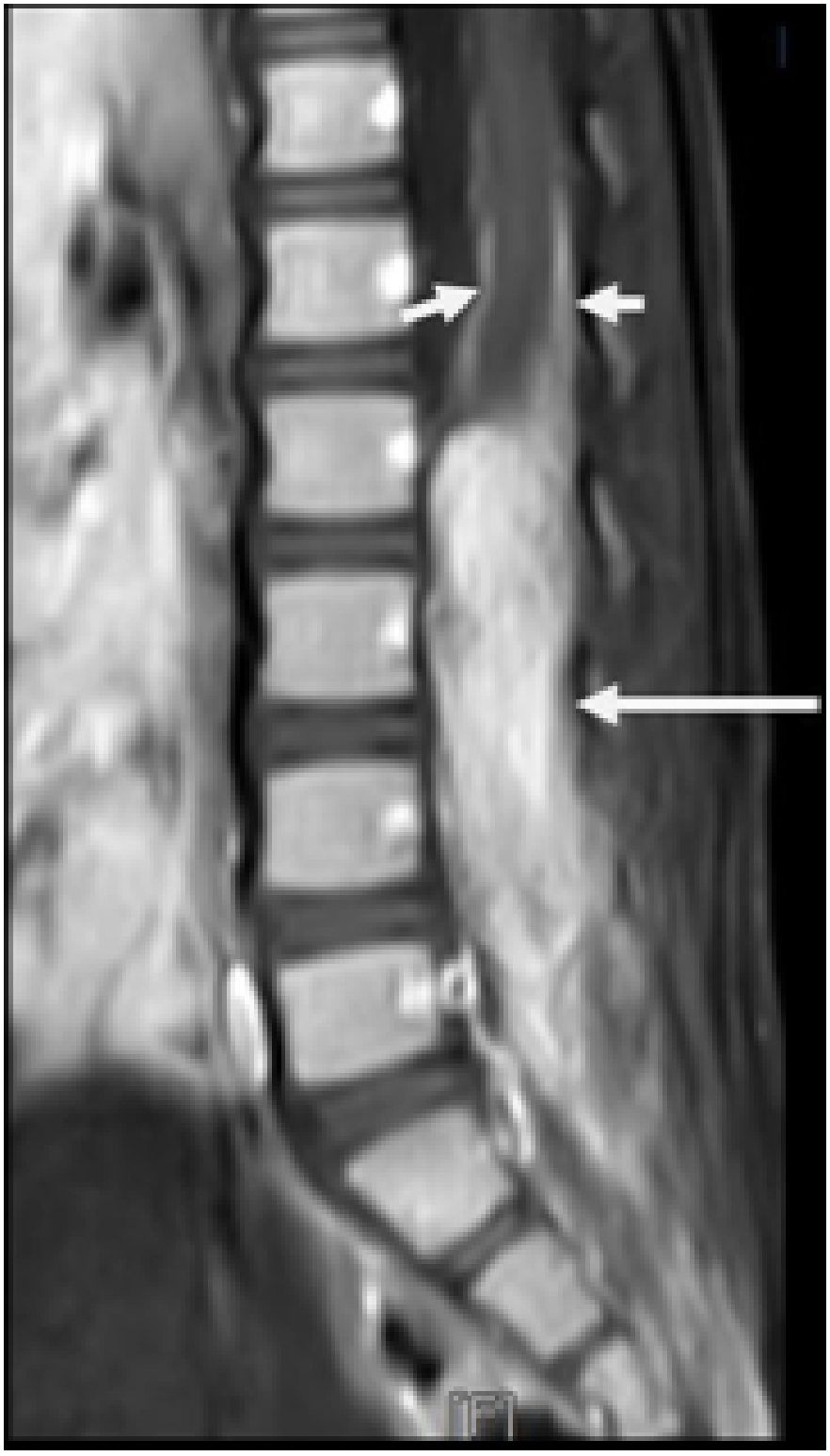
Moyson F, Maurus-Desmarez R, Gompel C. [Mediastinal Wilms' tumor?] Acta Chir Belg.1961 Suppl 2:118–28.

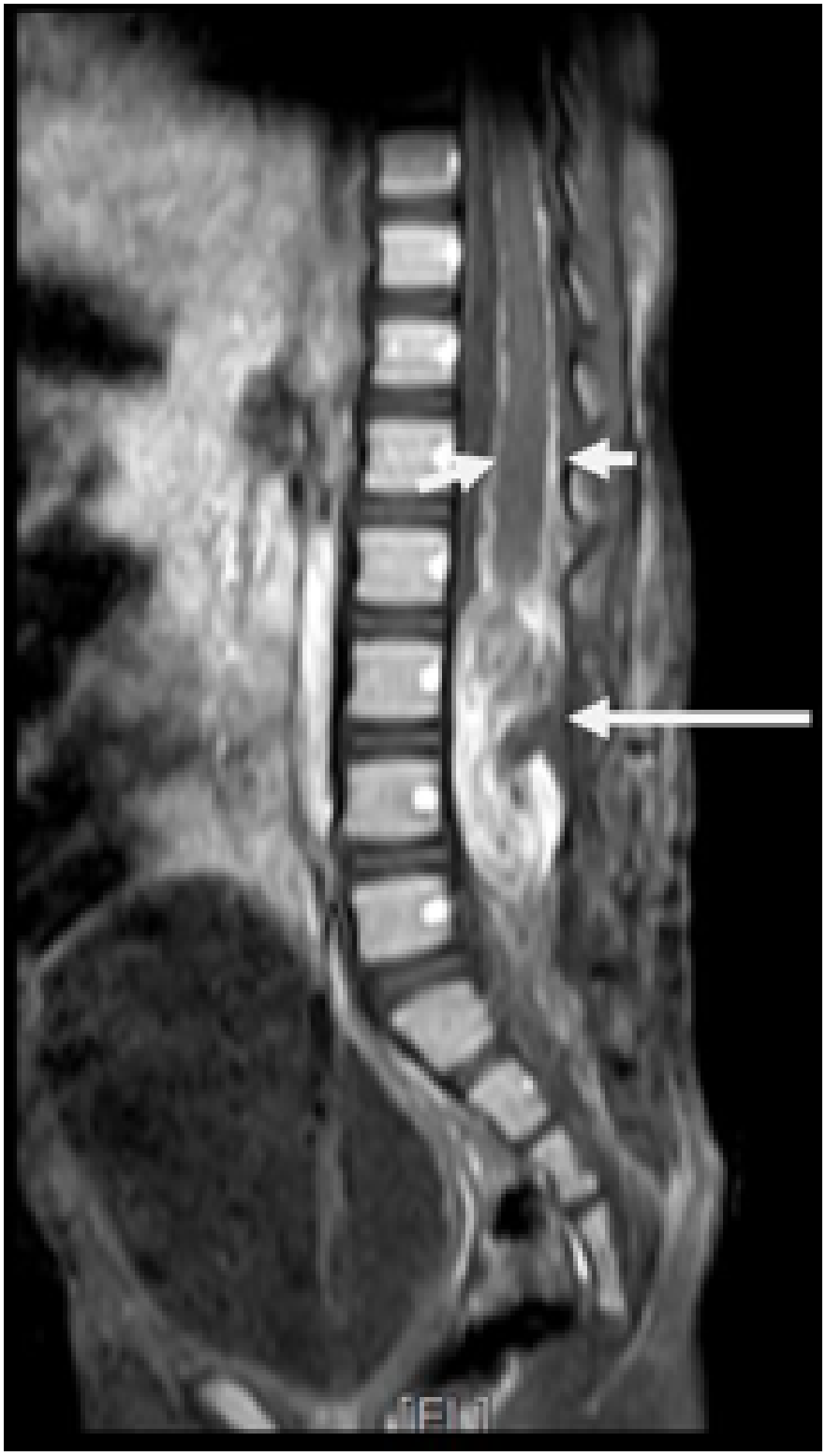
1. Armanda V, Culić S, Pogorelić Z, Kuljiš D, Budimir D, Kuzmić-Prusac I. Rare localization of extrarenal nephroblastoma in 1-month-old female infant. J Pediatr Urol.2012 Aug;8(4):e43–5.
2. Song JS, Kim IK, Kim YM, Khang SK, Kim KR, Lee Y. Extrarenal teratoid Wilms' tumor: two cases in unusual locations, one associated with elevated serum AFP. Pathol Int.2010 Jan;60(1):35–41.
3. Wu Y, Zhu X, Wang X, Wang H, Cao X, Wang J. Extrarenal nephroblastomatosis in children: a report of two cases.BMC Pediatr.2014 Oct 6;14:255.
4. Andrews PE, Kelalis PP, Haase GM. Extrarenal Wilms' tumor: results of the National Wilms' Tumor Study. J Pediatr Surg. 1992;27:1181-1184.
5. Mickle JP, McLennan JE. Malignant teratoma arising within a lipomeningocele. Case report. J Neurosurg. 1975;43:761-763.
6. Sharma MC, Jain D, Sarkar C, Bhatnagar V, Rishi A, Suri V, et al. Lumbosacral Wilms' tumor as a component of immature teratoma associated with spinal dysraphism: a rare case and short literature review. Fetal Pediatr Pathol. 2009;28:201-208.
7. Song JS, Kim IK, Kim YM, Khang SK, Kim KR, Lee Y. Extrarenal teratoid Wilms' tumor: two cases in unusual locations, one associated with elevated serum AFP. Pathol Int. 2010;60:35-41.
8. Tebbi K, Ragab AH, Ternberg JL, Vietti TJ. An extrarenal Wilms' tumor arising from a sacrococcygeal teratoma. Clin Pediatr (Phila). 1974;13:1019-1021
9. Sharma MC, Aggarwal M, Ralte AM, Vaishya V, Suri A, Gupta V, et al. Clinicopathological study of spinal teratomas: a series of 10 cases. J Neurosurg Sci. 2003;47:95-100.
10. Posalaky Z, Drake RM, Mawk JR, Franciosi RA, Spencer BH Athinarayanan PR, et al. Ectopic immature renal tissue over the dorsum of lumbar and sacral area in two infants. Pediatrics. 1982;69: 336-339.
11. Fahner JB, Switzer R, Freyer DR, Mann JD,Mann RJ. Extrarenal Wilms' tumor. Unusual presentation in the lumbosacral region. Am J Pediatr Hematol Oncol. 1993;15:117-119.
12. Ratajczak MZ, Schneider G, Zachariah PS, Kucia M, Kakar SS. The embryonic rest hypothesis of cancer development: an old XIX century theory revisited. J Cancer Stem Cell Res. 2014;2:e1001.
13. . Sell S. On the stem cell origin of cancer. Am J Pathol. 2010;176:2584-2594.

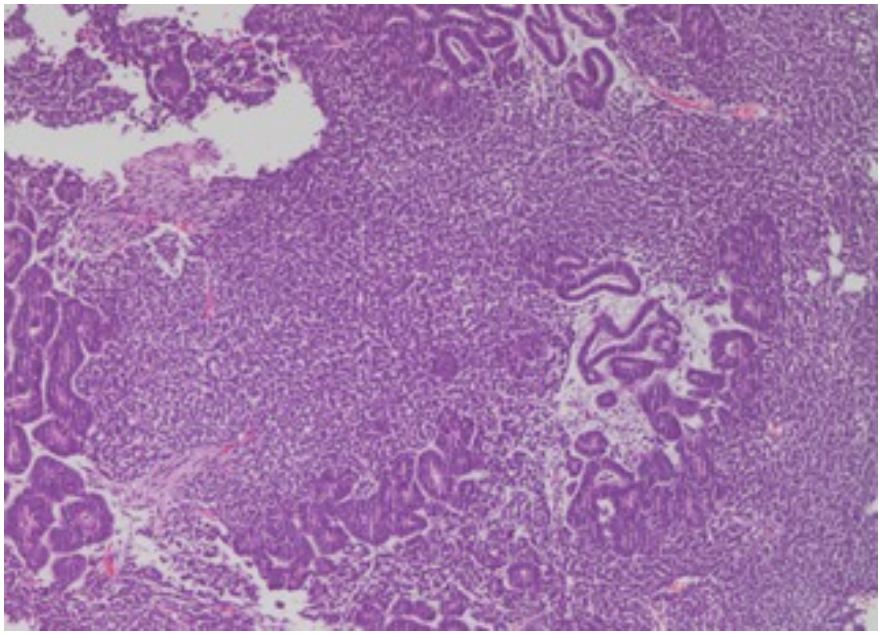
14. Fernbach SK, Naidich TP, McLone DG,Leestma JE. Computed tomography of primary intrathecal Wilms tumor with diastematomyelia.J Comput Assist Tomogr. 1984;8:523-528.
15. Grobstein C. Inductive tissue interaction in development. Adv Cancer Res. 1956;4:187-236.
16. Deshpande AV, Gawali JS, Sanghani HH,Shenoy AS, Patankar JZ, Borwankar SS. Extrarenal Wilms' tumour: a rare entity. Pediatr Surg Int. 2002; 18:543-544.
17. Horenstein MG, Manci EA, Walker AB, Dehner LP. Lumbosacral ectopic nephrogenic rest unassociated with spinal dysraphism. Am J Surg Pathol. 2004;28:1389-1392..
18. Coppes MJ, Wilson PC, Weitzman S. Extrarenal Wilms' tumor: staging, treatment, and prognosis. J Clin Oncol. 1991;9:167-174.
19. Sastri J, Dedhia R, Laskar S, Shet T, Kurkure P, Muckaden M. Extra-renal Wilms' tumoureis it different? Pediatr Nephrol. 2006;21:591-596.











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