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Case Report

An Astonishing Extrarenal Wilms Localisation; Spinal Cord

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ABSTRACT

Wilms' tumour is a renal tumour mostly seen during the first 5 years of life and it accounts for 95% of renal malignancies during childhood. Its origin is primitive metanephric cells and, very rarely, it may occur in places other than the kidneys. The estimated rate of nephroblastoma outside the kidneys is approximately 0.5 to 1% of Wilms' tumour cases. In this article, we report on a 3-year-old female patient 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 tumour arose in this location. This is a very rare extrarenal Wilms' tumour location. Here, we report on a case with immature renal cells located in the lumber spinal cord associated with spinal dysraphism and the development of Wilms' tumour there after one year.

Keywords: Children, Extrarenal, Wilms

Introduction

Wilms' tumour is a renal tumour mostly seen during first 5 years of life and it accounts 95% of renal malignancies during childhood. Its origin is primitive metanephric cells and, very rarely, it may occur in places other than the kidneys. The estimated rate of nephroblastoma outside the kidneys is approximately 0.5 to 1% of Wilms' tumour cases.

Wilms' tumour is one of the most common childhood solid malign tumours, mostly diagnosed during the first five years of life. 95% of renal malignancies are known to be Wilms' tumours in paediatrics, and they arise from primitive metanephric cells.

Very rarely, this tumour may occur in places other than the kidneys. Wilms' tumour located outside the kidneys without any primary tumour in the kidneys is called an extrarenal Wilms' tumour (ERWT). ERWT was first described by Moyson et al. (1) in 1961. The estimated ERWT rate is approximately 0.5 to 1% of Wilms' tumour cases. The prognosis is similar to that of renal Wilms' tumour. ERWT occurs mostly in childhood and rarely in adults (2).

Wilms' tumour may be observed outside the kidneys in two other situations: metastatic disease and Wilms' tumour arising in a teratoma. Therefore, for a diagnosis of ERWT, a metastatic lesion or teratoma should be ruled out. For this reason, it is necessary to evaluate the kidneys for primary tumour preoperatively and search the whole specimen for any teratoid element postoperatively (3).

In renal Wilms' tumours, it is known that, when the persistent intrarenal fetal nephrogenic blastemal tissue undergo oncogenic change, a tumour appears. It is thought

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that, in ERWT cases, the ectopic nephrogenic rests may develop into a Wilms' tumour with the same mechanism. Several reports have described nephrogenic rests in the inguinal, retroperitoneal and lumbosacral regions (4).

In this article, we report a 3-year-old female patient 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' tumour arose in this location. ERWT located in the spine is very rare. Thus, we report this case of a congenital Wilms' tumour associated with spinal dysraphism to increase awareness.

Case Report

A 3-year-old girl was admitted to our clinic with difficulty in walking beginning 1.5 months prior to admission. We learned that she had been operated on five times beginning at two months of age with dermal sinus and tethered cord diagnoses. However, no pathological examination was carried out on the operation specimen.

1.5 months previously, 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 magnetic resonance imaging (MRI) (Figure 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 was blastemal nodules. The proliferation index was 2.8%, and mitosis were rare (Figure 2).

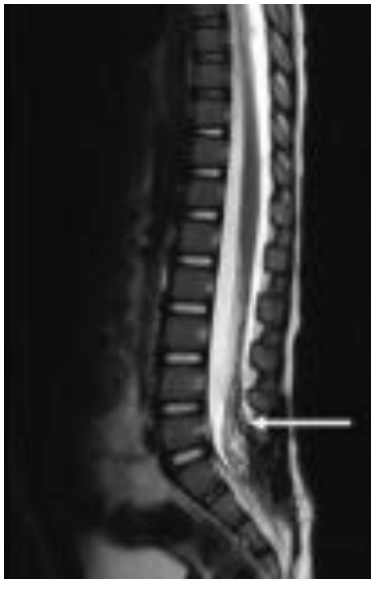

Figure 1. T2-weighted sagittal lumbosacral MR image shows a mass lesion within spinal canal between L2-S2 level (arrow)

After 1.5 months, the patient was readmitted with complaints of increased difficulty in walking, standing, and balance.

The patient was hospitalized. She had difficulty in walking, and she was incontinent for 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. With these 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 (Figure 3). Urinalysis, renal, and liver function tests were normal. Abdominal ultrasonography and MRI were normal. Surgical resection of the tumour was

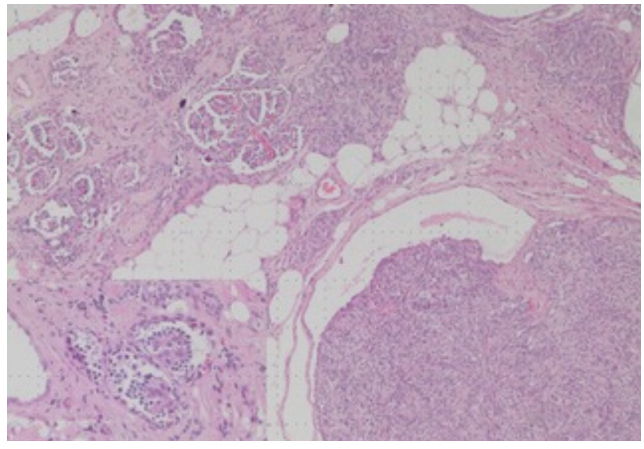

Figure 2. The lesion shows multiple immature glomeruli and tubules (upper left) and blastemal nodule (lower right) along with intervening fat tissue (H&E, X10). Immature glomeruli (H&E, X40, inside)

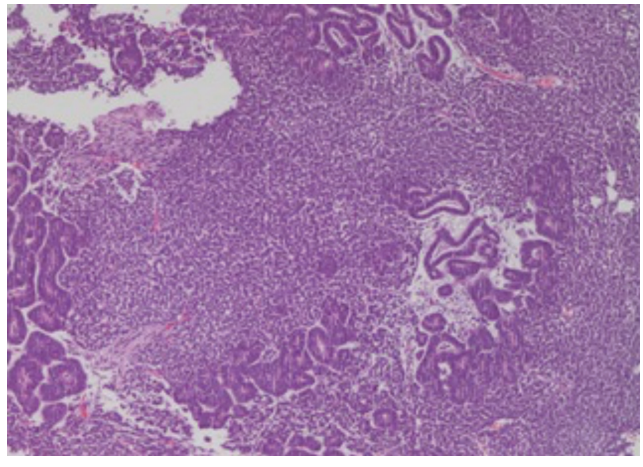

Figure 3. Contrast enhanced sagittal T1-weighted lumbosacral MR image shows the mass is enlarged, extending from L2 to S2 level (Arrow). The mass enhances markedly and there is also leptomeningeal enhancement around distal spinal cord (short arrows)

MR: Magnetic resonance

performed, but the tumour mass could not be completely removed (Figure 4). The histopathological examination of the mass revealed triphasic histology Wilms' tumour in which blastemal, stromal, and epithelial elements were present (favourable histology) (Figure 5). According to the National Wilms' Study Group protocol, the tumour was accepted as Stage III. She completed her chemotherapy and radiotherapy. She is being followed up in the oncology outpatient clinic. Physical examination reveals weakness in her right lower extremity (muscle strength 4/5). She has no incontinence.

Discussion

ERWT located at spinal cord is very rare. Wilms' tumour occurring within teratomas is another entity and it is diagnosed as teratoid Wilms' tumours pathologically (5-10). No elements of teratoma were found in our patient's specimen.

Posalaky et al. (11) described nephrogenic rests in the spine in two cases. There was no associated spinal dysraphism and the nephrogenic rests were benign with no evidence of ERWT. However, ten years later, Fahner et al. (12) described ERWT in the spine.

It is postulated that embryonic rests cause spinal dysraphism and ERWT (13). Some changes in the tissues around of these rests may cause them to proliferate and form masses that are like fetal tissues (14). Thus, according to this theory, ERWT arises from pluripotent mesenchymal cell rests which may transform into malignant masses (13,14). Fernbach et al. (15), and Grobstein (16) suggested that the embryonic central nervous system may induce nephrogenic differentiation in the embryonic mesenchyme from which the spine develops. Deshpande et al. (17) reported an ERWT within the dorsal lumbar spine's subcutaneous fat, and Horenstein et al. (18) reported nephrogenic rests in the same location.

The nephrogenic remnants in the spine, outside the kidneys, as in our case, supports the hypothesis that mesenchymal cells in the wrong place may develop into ERWT after an unknown stimulation (13).

In a review of 34 cases of ERWT, Coppes et al. (19) suggested that patients with ERWT receive postoperative chemotherapy, with the same protocol as for a renal Wilms' tumour. Sastri et al. (20) reviewed three additional cases and summarized a total of 48 cases of ERWT, and was in agreement with Coppes et al. (19).

Our patient was treated according to the latest guidelines for renal Wilms' tumour, following the National Wilms Tumour Study-V regimen for EE4A.

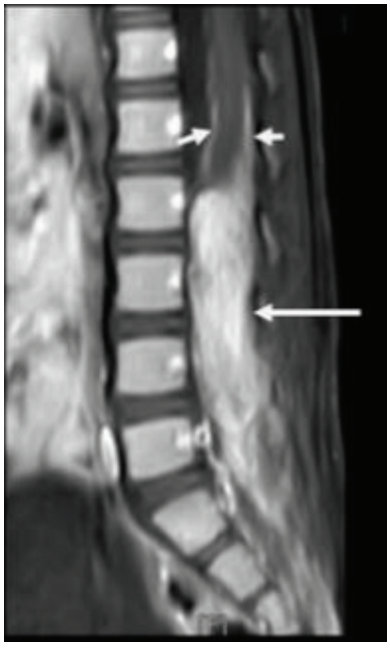

Figure 4. Contrast enhanced sagittal T1-weighted lumbosacral MR image shows post-operative defect within the mass lesion (arrow). The mass is slightly smaller than the previous MR examination. Leptomeningeal enhancement is still present (short arrows)

MR: Magnetic resonance

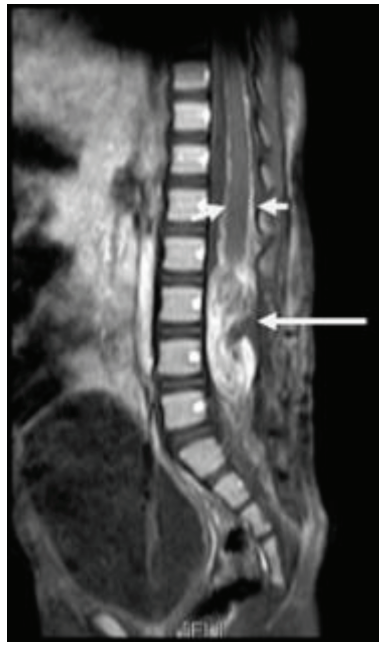

Figure 5. Diffuse blastemal component with small to medium-sized undifferentiated cells and epithelial component with tubule-like structures (H&E, X10)

Conclusion

In this article, we report a 3-year-old female patient 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' tumour arose in this location. ERWT located in the spine is very rare. This report may explain the pathogenesis of ERWT with the embryonic rest theory.

Ethics

Informed Consent: Written informed consent was obtained from the patient's parents.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Concept: A.G.T., N.E., B.T.T., S.U.B., M.S., S.A., K.E.A., Design: A.G.T., N.E., B.T.T., S.U.B., M.S., S.A., K.E.A., Data Collection and/or Processing: A.G.T., N.E., B.T.T., S.U.B., M.S., S.A., K.E.A., Analysis and/or Interpretation: A.G.T., N.E., B.T.T., S.U.B., M.S., S.A., K.E.A., Literature Search: A.G.T., N.E., B.T.T., S.U.B., M.S., S.A., K.E.A., Writing: A.G.T., N.E., B.T.T., S.U.B., M.S., S.A., K.E.A.

Conflict of Interest: The authors declared that there were no conflicts of interest.

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