CASE REPORT

A case of Wilms' tumor with spinal cord involvement

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Abstract

Wilms' tumor is the most common renal tumor of childhood. However, spinal cord involvement has rarely been reported in children with Wilms' tumor. In this article, we report a 5-year-old boy with Wilms' tumor who had the unusual presentation of spinal cord involvement. Our purpose is to emphasize that spinal cord involvement may be seen in children with Wilms' tumor. (J Pediatr Neurol 2003; 1(1): 47-50).

Key words: Wilms' tumor, spinal cord, child.

Introduction

Wilms' tumor is the most common renal tumor of childhood and presents as an abdominal mass with pain, hematuria, hypertension, fever, anorexia, and weight loss (1). Wilms' tumor has an incidence of metastasis of approximately 10% (2). Lymph nodes, lung, liver, and bone are the most common sites of metastatic disease, whereas intraspinal metastasis is rare (3-6). Epidural involvement usually occurs with other evidence of spreading (3-6). Spinal cord metastasis without concomitant involvement to other organs is extremely rare (4,7). Herein, we report a case of Wilms' tumor with spinal cord involvement but without other organ metastasis.

Case Report

A 5-year-old boy was admitted to our clinic with lumbar pain beginning 2 months before admission. A generalized afebrile seizure lasting about 5 minutes was noted in our emergency room. On physical examination high blood pressure (150/100 mm/Hg) was noted. He also had a left sided abdominal mass, which was 13x10 cm in diameter. He had no meningeal signs and the remainder of the physical findings were unremarkable. It was presumed that his seizure was probably due to hypertension. The admitting physician did not evaluate muscle strength and deep tendon reflexes. The child, however, could walk and run.

Urinalysis, renal and liver function tests were normal except for hematuria. Cerebrospinal fluid examination was also normal. Abdominal ultrasonography showed anterior and inferior displacement of the left kidney due to an echogenic mass 11 x 8.5 cm in diameter. Intravenous urography also showed a left sided renal mass with a normal right kidney. Computerized tomography (CT) of the abdomen confirmed the mass in the left kidney and revealed multiple lymph nodes around the left renal hilus without evidence of inferior vena cava thrombus. Chest and brain CT were normal. The histopathological examination of the mass revealed triphasic histology Wilms' tumor in which blastemal, stromal, and epithelial elements were present (favorable histology). According to the National Wilms' Study Group protocol the tumor was accepted as Stage II-B.

The patient was hospitalized and immediately and exploratory laparotomy was performed. A firm well-encapsulated and lobulated renal tumor was completely excised and samples from the lymph nodes around the renal pelvis were taken. The contralateral kidney was normal. After the operation, the patient had difficulty walking and he was incontinent of urine and stool. Physical examination revealed weakness in the lower extremities (muscle strength was 1/5 bilaterally).

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Figures 1 and 2. Lumbar magnetic resonance imaging shows the mass compressing the spinal cord and involving vertebra at the L2 level.

and deep tendon reflexes were absent. Sensory examination of touch, pain and temperature were unremarkable. 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 level compressing the spinal cord (Figures 1 and 2). Surgical resection of the tumor was performed but the tumor mass could not be completely removed. The patient is 2 months follow in surgery. Physical examination reveals weakness in the right lower extremity (muscle strength 3/5), left lower extremity (muscle strength 4/5) and deep tendon reflexes are absent. Urinary and fecal incontinence did not recover. He is being given adjuvant chemotherapy and radiation therapy.

Discussion

Wilms' tumor, an embryonal renal neoplasm, has an incidence of metastasis of approximately 10% (2). Lymph nodes, lung, liver and bone are the most common sites of metastatic disease, whereas intraspinal metastasis is rare (3-6). Several different mechanisms are implicated in the pathogenesis of spinal cord metastasis, including direct extension from a paravertebral tumor mass, hematogenous spreading through collateral circulation of the paravertebral venous plexus, lymphatic spreading through the vertebral foramen, or spreading along the perineurium of the spinal nerves (8). We think there was direct extension of the tumor to the vertebral body and spinal cord in our case.

Patients with Wilms' tumor with spinal

metastases with or without other organ involvement have only rarely been reported in the literature (Table 1) (3-7,9-14). Additionally, Arrotegui et al. (15) reported a further case of Wilms' tumor associated with spinal cord involvement. Our patient had spinal involvement without evidence of lung or liver metastasis. Although bone involvement is relatively frequent in patients with sarcomatous Wilms' tumor, our patient had vertebral involvement despite a favorable histology (16).

The particular constellation of symptoms that accompanies metastatic epidural cord compression varies with the location of the lesion. Localized back pain in the area of the lesion is the most common symptom of spinal metastasis and may be the only symptom of an epidural lesion for 2-7 months before the emergence of other symptoms of spinal cord compression (17). In addition to back pain, weakness, sensory loss with gait disturbance and autonomic dysfunction may be seen. Rarely, intraspinal tumors may present with abdominal pain as the initial symptom (18). Our patient presented with back pain and then inability to walk. It is imperative that early signs of cord compression be recognized, because prompt diagnosis and treatment are mandatory to prevent irreversible loss of neurologic function (7).

Authors have reported that clinically aggressive Wilms' tumor and poor outcomes are associated with metastases to sites other than the lung, and in agreement with this observation, the mortality rate of patients with intraspinal metastases is high (7,19). Children with intraspinal metastatic Wilms' tumor however can achieve long-term disease free survival with intensive multimodality therapy.

Table 1. Patients with Wilms' tumor with intraspinal metastases

References	Patient no	Age at diagnosis	Tumor stage	Histology of primary tumor	Time to spinal metastasis	Presenting symptoms of cord compression	Other sites of metastatic disease	Treatment of spinal metastasis	Outcome
Ebb et al. (5)	1	5 yr 6 mo	IV	Anaplastic	NR	Back pain	NR	S, RT	Dead
Ebb et al. (5)	2	3 yr 6 mo	IV	Anaplastic	16 mo	LE weakness	NR	S, RT	Alive with disease
Bever et al. (3)	3	4 yr	II	Anaplastic	9 mo	Back pain	Lung	S, L, SR	Alive with disease
Bever et al. (3)	4	6 yr	I	NR	14 mo	Back pain	Lung, adjacent vertebra	SR, RT, CT	NED
Sumner et al. (4)	5	7 yr	NR	Sarcomoid foci	6 mo	LE weakness	Adjacent vertebra	L, RT	Dead
Chang et al. (10)	6	4 yr	I	NR	12 mo	LE pain	None	S, RT, L, SR	NED
Dickerhoff et al. (6)	7	5 yr	NR	NR	0*	Paraparesis	None	L, CT, RT	Dead
Dickerhoff et al. (6)	8	14 yr	NR	NR	9 yr	Back pain	Lung	CT	Alive with disease
Keidan (11)	9	2 yr	NR	NR	6 mo	Abdominal and back pain	Retroocular mass, bone	RT	Dead
Kim et al. (12)	10	1 yr 2 mo	I	Favorable	7 mo	Paraplegia	Lung	L, SR, RT, CT	Dead
Rey et al. (13)	11	3 yr 3 mo	I	Favorable	7 yr 2 mo	LE weakness and back pain	Lung	None	Dead
Wiegel et al. (14)	12	9 yr	I	Anaplastic	8 mo	Back pain	None	SR, RT	NED
Cohn et al. (7)	13	4 yr	II	Favorable	11 mo	Abdominal pain	Adjacent vertebra	RT, CT	NED
Cohn et al. (7)	14	8 yr	IV	Favorable	16 mo	Abdominal and side pain	None	L, SR, RT, CT	NED
Darendeliler et al. (9)	16	5 yr	IV	Anaplastic	0*	LE weakness	Abdominal	ABMT	Alive with disease
Presented patient	17	5 yr	IV	Favorable	0*	Paraplegia	Abdominal	SR, CT, RT	Alive with disease

NR: Not reported; S: Steroid; RT: Radiation therapy; CT: Chemotherapy; L: Laminectomy; SR: Surgical resection; ABMT: Autologous bone marrow transplantation; NED: No evidence of disease; LE: Lower extremity; EXP: Experimental treatment (ICRF 159); CCSK: Clear cell sarcoma; NVVTS: National Wilms' Tumor Study Registry.
*Reported with cord metastasis at diagnosis.

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