Case Report

A Wilms’ Tumor with Spinal Cord Compression: An Extrarenal Origin?

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Spinal cord compression in Wilms’ tumor (WT) is an extremely rare event that can have a very poor prognosis if not taken care of rapidly. Most cases reported in the literature involve widely metastatic patient with bone or paraspinal metastases or occasionally intradural metastasis. Here, we present the case of a 3-year-old girl of WT confirmed by biopsy, with spinal cord compression due to the direct contiguous spread of a tumor through 2 vertebral foramina. Abdominal ultrasonography and magnetic resonance imaging performed for an abdominal mass revealed a large heterogeneous tumor near the upper pole of the left kidney. An nodular infiltration extended through the T11-L1 and L1-L2 neural foramina, forming an intraspinal mass that compressed the spinal cord. Major paresthesia subsequently occurred, requiring urgent treatment with corticosteroids and chemotherapy. The evolution was rapidly satisfying. After six courses of chemotherapy, a left nephrectomy was performed. Macroscopic examination identified a large tumor attached to the kidney without renal infiltration. Microscopic examination concluded to a nephroblastoma with regressive changes, of intermediate risk. Evolution at 6 months is satisfactory, with no neurological deficit. The histological aspect of the tumor and the clinical outcome suggest that she had an extrarenal WT that spread through the vertebral foramina and was secondarily attached to the kidney.

1. Introduction

Childhood renal neoplasm accounts for approximately 7% of all cancers in childhood and are in the vast majority Wilms’ tumor (WT) or nephroblastoma [1, 2]. About 10% of WT present with haematogenous spread, most commonly to the lungs (85%), liver (10%) and only very rarely to the bones (1%) and brain [1, 2]. The occurrence of spinal cord compression ranges from 2.7 to 4% in childhood neoplasm, generally in metastatic or invasive Ewing’s sarcoma, osteogenic sarcoma, rhabdomyosarcoma, neuroblastoma, and lymphoma [3, 4]. Spinal cord compression may result in permanent neurological deficit, further aggravating the burden of disease.

In the course of WT, spinal cord compression is a very rare occurrence, usually involving skeletal metastases to the vertebral body, intradural or extradural metastases [5–11].

Here, we report the case of a large WT in a 3-year-old patient with secondary spinal compression by direct contiguous spread through 2 vertebral foramina.

2. Case Presentation

A 3-year-old girl, with no prior medical history, was admitted in our center with a three-week history of an abdominal mass discovered by her mother. On physical examination, a firm, painless mass in the left flank was palpable. Complete examination showed no other abnormality. In particular, no neurological deficit was detected.

Abdominal ultrasonography revealed a large heterogeneous tumor of $69 \times 67 \times 97$ cm originating from the upper...
pole of the left kidney, deviating it towards the midline. The mass is located on the periphery of the upper pole of the kidney, and a vascular pedicle seemed to emerge from the renal sinus. No calcification or hemorrhagic component was found. Magnetic resonance imaging (MRI) and computed tomography (CT) showed an encapsulated tumor but with a nodular infiltration of the retroperitoneal fatty tissues. It extended through the T11-T12 and T12-L1 neural foramina, forming an intraspinal mass from T11 to L1 and compressing the spinal cord (Figure 1). Assessment of tumor extension revealed two infracentimetric metastases in the lungs. The tumor and its extradural extension showed a major hypermetabolic activity on positron emission tomography (PET). Bone marrow aspiration uncovered no medullary involvement. The urine catecholamines, neural specific enolase, alpha-fetoprotein, and human chorionic gonadotropin were normal. Laboratory studies evidenced only a small rise in LDH (417 IU/L) and fibrinogen (7.2 g/L).

Considering this extremely unusual clinicoradiological presentation, a posterior transcutaneous needle biopsy was performed, as recommended in the International Society of Pediatric Oncology renal study group SIOP-RTSG 2001 protocol. The histopathologic features revealed a triphasic nephroblastoma, without anaplastic feature.

Meanwhile, the patient started complaining of major paresthesia and leg pain, requiring urgent treatment with corticosteroids and chemotherapy. Due to the neurological threat and the lung nodules, chemotherapy according to the SIOP-RTSG 2001 for stage IV nephroblastoma was administered, including three drugs (vincristine, actinomycine D, and doxorubicine).

The patient’s evolution was rapidly satisfying, with the rapid and complete receding of neurological symptoms. The preoperative assessment, after four courses of chemotherapy, indicated a massive regression of the tumor volume by 53%, with measures of $67 \times 46 \times 77$ cm, and a complete disappearance of the intraspinal extension. The lung nodules were no longer detected on CT imaging.

After six courses of chemotherapy, a left nephrectomy was performed. Macroscopic examination identified a large tumor attached to the kidney, enclosed in a thick fibrous capsule. The microscopic examination concluded to a triphasic nephroblastoma with regressive changes, of intermediate risk with only small areas of blastema. The histology of the kidney was unremarkable without any nephrogenic rest. Postoperative treatment included 29 weeks of chemotherapy with the same three drugs. After 24 months of evolution, the child is in good health and has no neurological deficit.
Spinal cord compression is a very rare occurrence in WT, but it can have dramatic functional and vital consequences if not taken care of appropriately. Treatment comprises of immediate chemotherapy and corticosteroids to reduce the tumor size, followed by nephrectomy.

While the vast majority of WT cases with spinal cord compression are explained by metastasis to the spinal canal (bone, extradural or intradural metastasis), we report the first case of contiguous spreading from the primary tumor through the neural foramina in a child devoid of spinal dysraphism. This case could be explained by the extrarenal origin of the nephroblastoma.

**Conflicts of Interest**

The authors declare that they have no conflicts of interest.

**References**


