Brain Metastasis in Wilms' Tumor: a Case Report

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Abstract- Wilms' tumor is the most common abdominal tumor of childhood, and its cerebral metastasis is apparently very rare. The authors report an 18-month-old girl with Wilms' tumor and brain metastasis.

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Introduction

In children with solid tumors, brain metastasis is relatively uncommon (1), accounting for only 0.5-1.8% of all pediatric craniocerebral tumors (2). Prior to the introduction of effective chemotherapy, intracranial metastasis of Wilms' tumor has been reported as a postmortem finding in up to 13% of patients dying of metastatic disease (3). Although the occurrence of central nervous system (CNS) metastasis in other tumors typically is associated with a rapid deterioration and often associated with widely disseminated disease (4), CNS metastasis in Wilms' tumor may occur as an isolated event. A total of 23 patients with Wilms' tumor metastasizing to the CNS have been reported to date (5). Because of this relative rarity, information regarding incidence, the pattern of spread, management, and prognosis is limited. To better understand this entity, we report an 18 months old girl with Wilms 'tumor who developed intracranial metastasis.

Case Report

An 18-month-old girl underwent left nephrectomy for a stage III Wilms' tumor of favorable histopathology. Treatment after complete resection included Actinomycin D, Vincristine and Doxorubicin according to the SIOP (93-01/GPOH) protocol for stage III Wilms' tumor. She was referred to our hospital five months after initial diagnosis. CT scan of the abdomen showed a 32x38 mm heterogenous, mild enhancing solid mass in left para-aortic region continued from renal hilum level to the pelvic rim, in favor of tumor recurrence. She was also noted to have pulmonary metastasis. Triple chemotherapy with Ifosfamide, Carboplatin and Etoposide (ICE) (10) was administered which produced a rapid response with complete resolution of the pulmonary metastasis and resolution of the abdominal tumor.

After four months, while receiving ICE protocol chemotherapy and radiation therapy, she presented with signs of increased intracranial pressure. Cranial CT scan demonstrated a large tumor in the frontal region (Figure 1). A subsequent craniotomy was performed, and surgical resection was followed by chemotherapy.

Histopathology of a brain tumor was consistent with metastatic Wilms' tumor (Figure 2). After craniotomy, chemotherapy (ICE Regimen) was continued for five courses and on six months follow, she is still clinically stable.

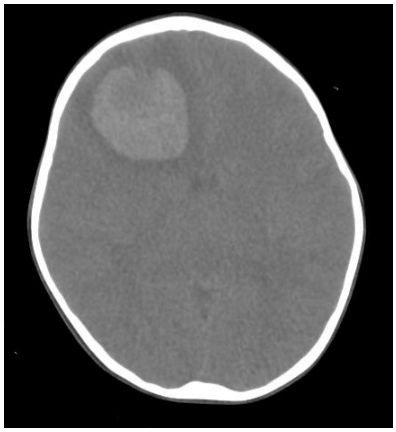

Figure1. Computed tomography scan: A large tumor in the right Frontal region

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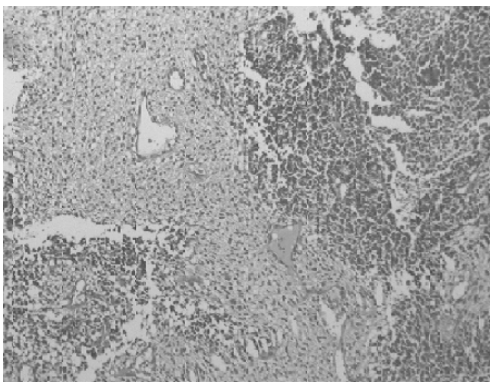

Figure 2. Clusters of primitive, undifferentiated tumor cells, as well as epithelial tubules, are seen in brain parenchyma. (H&E, X400)

Discussion

Brain metastases of solid tumors are rarely observed in children with cancer (4,6). Data regarding brain metastasis in pediatric solid tumor patients are limited, but clinical reports suggest that their frequency is 1.5-4.9% (1,4,6), and autopsy studies suggest a 6-13% frequency (7, 3). Brain metastases may be present at the initial diagnosis but in most cases, they develop later, during disease progression or relapse (8).

With the exception of the osteosarcoma, study of St.Jude Children's Research Hospital, the median survival of children who develop hematogenous spread to the brain is less than six months (9). It is possible that if detected earlier, before the onset of symptoms, the tumor burden in the parenchymal brain may be less and easier to manage (1).

Imaging studies that are useful for brain metastases detection include CT and MRI. Contrast-enhanced MRI detects two to three times as many lesions smaller than five mm in diameter (8).

The optimal treatment for patients with brain metastases depends on the tumor type, the number of brain lesions and the presence of other systemic metastases. For patients with solitary metastases and no systemic disease, surgery followed by radiotherapy and/or chemotherapy may be the best treatment (4, 6). For patients with multiple brain metastases, chemotherapy and radiotherapy only, without surgery may be of value (7).

In the present case, brain metastasis occurred nine months after the initial diagnosis of Wilms' tumor.

As in current patient, the development of a CNS metastasis appears to be associated most commonly concurrently or prior to lung metastasis (5).

As it was solitary metastases, craniotomy followed by chemotherapy seemed to be the best treatment. Current patient, in spite of brain metastasis, has survived for more than six months since the onset of cerebral metastasis.

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