

Isolated central nervous system metastasis in pediatric Wilms' tumor: A case report and review of literature



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Introduction

In children with solid tumors, brain metastasis is relatively uncommon, accounting for only 0.5–1.8% of all pediatric craniocerebral tumors. Prior to the introduction of effective chemotherapy, intracranial metastasis of Wilms' tumor has been reported as a post-mortem finding in up to 13% of patients dying of metastatic disease. 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, 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. 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.

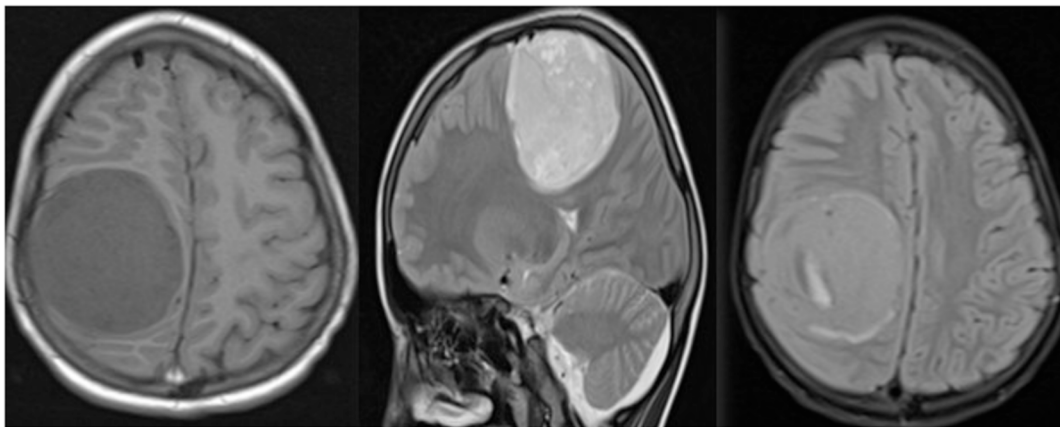


Fig. 1. Hypodense mass in the right fronto-parietal region with very little edema.

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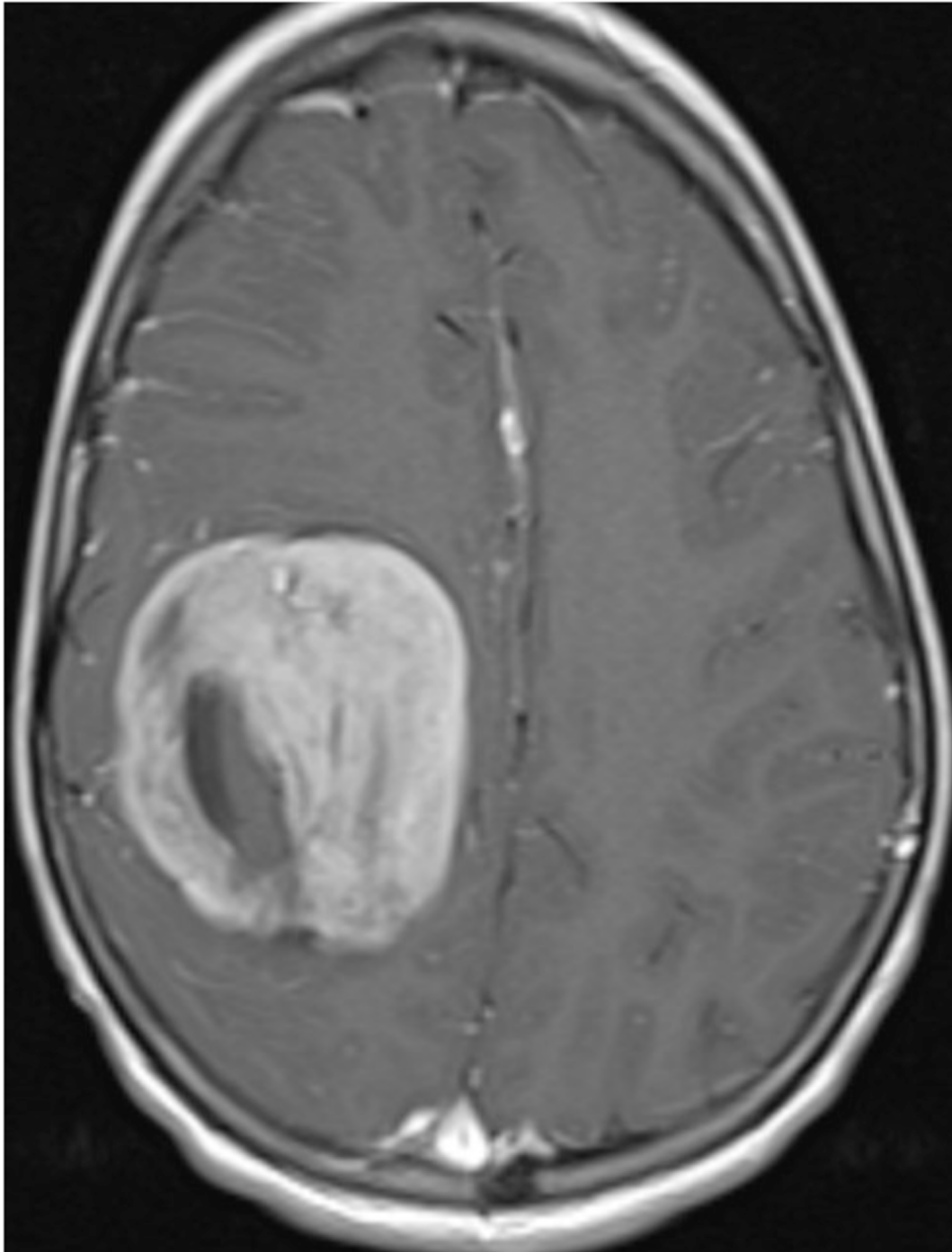


Fig. 2. Marked homogenous enhancement of the lesion.

Case report

A 10-year-old boy underwent left nephrectomy for Wilms' tumor with pulmonary and hepatic metastasis. Treatment after complete resection of the tumor included chemotherapy and radiotherapy. He was referred to our Neurosurgery department six months after initial diagnosis. He complained of frontal headache of increasing severity associated with weightiness in the left hemibody. On admission, his state of consciousness was found to be decreased and he had a left hemiplegia. A cranial computed tomography scan and a cerebral MRI were performed immediately showed an hypodense mass in the right frontoparietal region with very little edema (Fig. 1). After an intravenous injection of contrast medium (Fig. 2) there was marked homogenous

enhancement of the lesion. A subsequent craniotomy was performed with removal of the neoplasm. Histopathological examination (Fig. 3) confirmed that it was a metastatic Wilms' tumor. After the operation, the patient was treated with radiotherapy directed to the whole brain, and additional courses of chemotherapy were given. On a follow up examination 4 months later, the patient remained clinically stable with partial regression of his deficit and no other associated localization signs.

Discussion

Brain metastases of solid tumors are rarely observed in children with cancer.¹ Data regarding brain metastasis in pediatric solid tumor

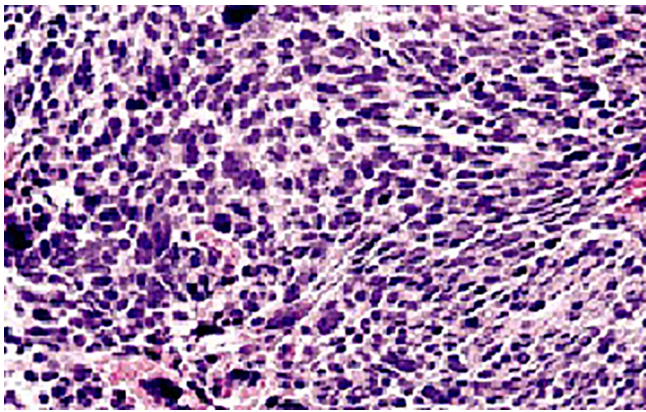


Fig. 3. Photomicrographs of histological sections demonstrating blastemal components in the brain metastasis.

patients are limited, but clinical reports suggest that their frequency is 1.5–4.9%,^{1,2} and autopsy studies suggest a 6–13% frequency.² Brain metastases may be present at the initial diagnosis but in most cases, they develop later, during disease progression or relapse.³ 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. 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.⁴ 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.³ For patients with multiple brain metastases, chemotherapy and radiotherapy only, without surgery may be of value. In the present case, brain metastasis

occurred six 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.

Conclusion

Wilms tumor is the most common renal malignancy of childhood and the second most common extracranial solid neoplasm occurring in children. Brain metastases are rare in the natural history of Wilms tumor.

A far more common site of tumor metastases is the lung. Overall survival tends to be good in children with favorable histology, even with widespread disease.

Conflicts of interest

The authors declare that there are no conflicts of interest regarding the publication of this article.

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