LUNG METASTASES IN NEUROBLASTOMA AT INITIAL DIAGNOSIS: a case report

M. Belcheva, Hr. Hristozova, V. Kaleva

Pediatric clinic of Oncohematology, University Hospital St Marina, Varna, Bulgaria

Introduction

Neuroblastoma is the most common extracranial pediatric solid tumor. Nearly 60% of all patients have metastatic disease at diagnosis. Bone and bone marrow are the most common sites of metastasis. Lung metastasis is uncommon and usually attributed to an advanced disease. Initial pulmonary metastases are extremely rare in neuroblastoma (0.7 up to 3.6% of stage IV patients) and are seen in tumors with aggressive phenotype.

Case presentation

A 16-month-old girl presented to a tertiary pediatric center with fever, cough, progressive dyspnea, malaise, weight loss and severe anemia. Chest radiographs revealed persistent bilateral pulmonary opacities (Fig.1). The child was initially diagnosed with pneumonia and an antibacterial treatment was started without efficacy. Cystic fibrosis and pulmonary tuberculosis were then ruled out. A subsequent chest CT scan revealed multiple pulmonary lesions between 5 and 25 mm in diameter, with smooth and spiculated margins, located predominantly in the lung bases; hilar adenopathies and pleural thickening next to pleural-based nodules (Figs. 2, 3). An abdominal CT scan revealed a large retroperitoneal mass thought to arise from the superior pole of the left kidney (Fig. 4). The child was then diagnosed with stage IV Wilms' tumor and underwent a treatment according to the nephroblastoma protocol. No measurement of urine catecholamines, nor histologic examination were undertaken. After 3 cycles of chemotherapy (Vincristin, Etoposide, Farmorubicin) a partial response of the primary tumor and no response at the metastatic sites was documented. The abdominal tumor was deemed inoperable at this moment and the parents declined subsequent treatment.

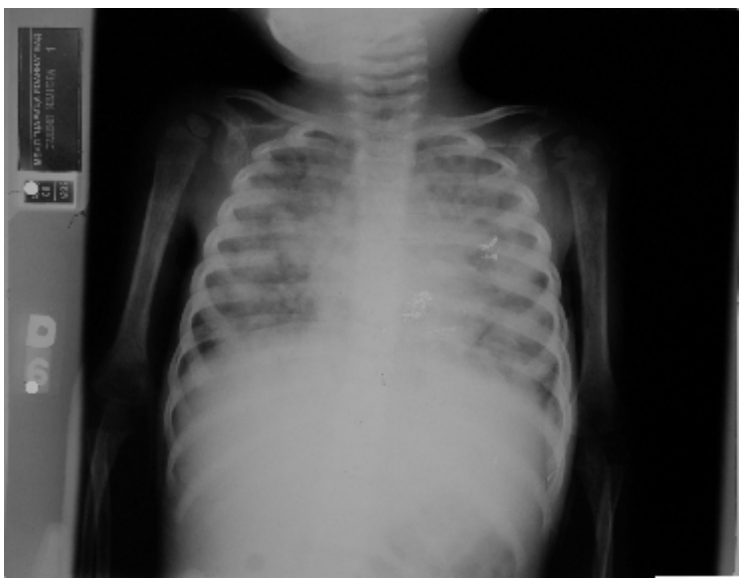
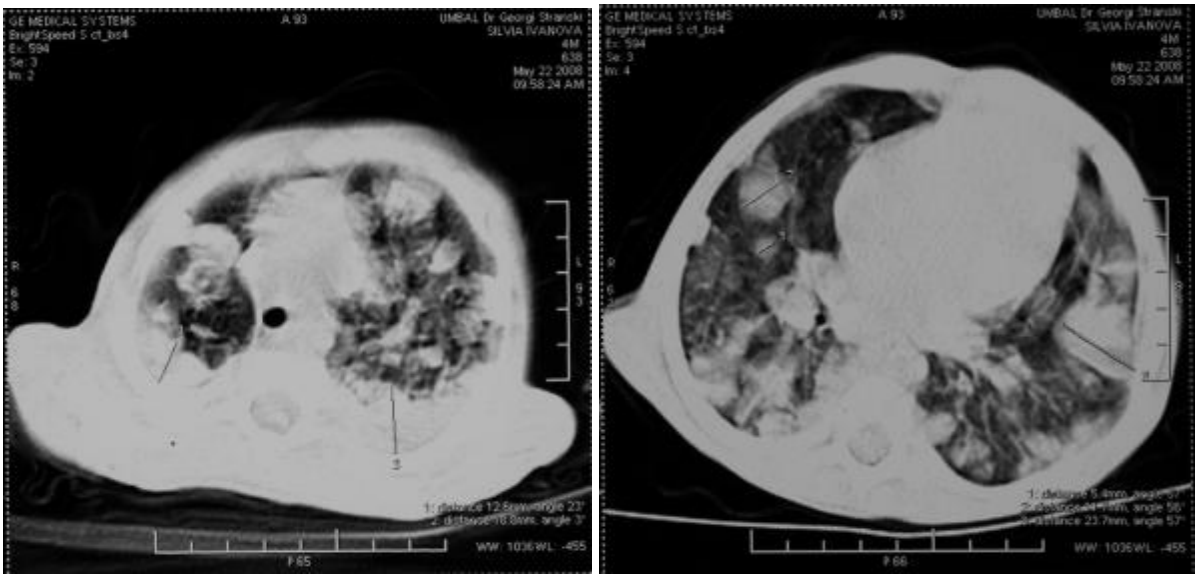


Fig.1. Initial chest X-ray showing diffuse bilateral pulmonary opacities



Figs.2, 3. Chest CT scan: multiple pulmonary lesions with smooth and spiculated margins; hilar adenopathies; pleural thickening next to pleural-based nodules

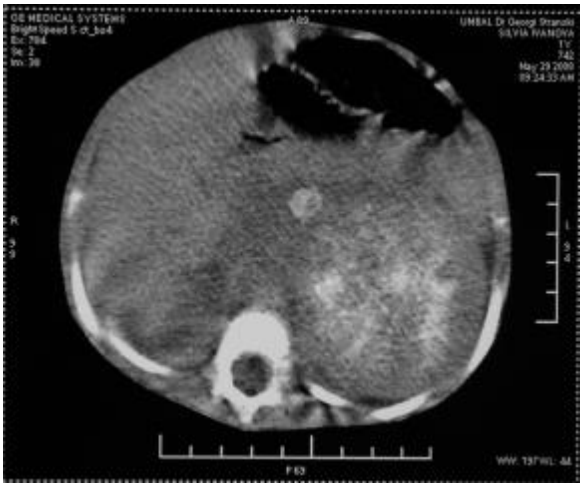
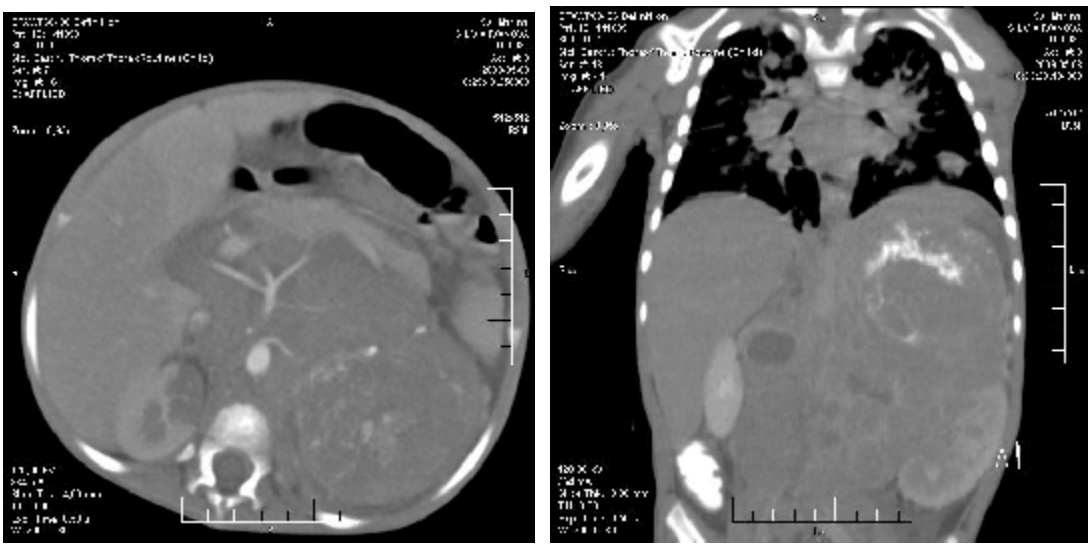


Fig. 4. Abdominal CT scan at diagnosis showing a large retroperitoneal mass displacing IVC

Nine months after therapy had been discontinued the child was admitted to our institution with a huge abdominal mass and severe anemia. There was marked arterial hypertension and no secretory diarrhea. The radiological examination revealed a large abdominal mass with calcifications, infiltrating the left kidney (Figs.5**,** 6) with extension around the large abdominal vessels, as well as multiple abdominal adenopathies and multiple bilateral pulmonary infiltrates and nodules. There were no CNS metastases. The LDH and ferritine levels exceeded 10 times the normal values. The histologic examination (Tru-cut and bone marrow biopsy) confirmed an unfavorable histology neuroblastoma with bone marrow dissemination.



Figs. 5, 6. Abdominal CT scan and CT scan with reconstruction

Discussion

Pulmonary metastases were the initial clinical presentation of left adrenal neuroblastoma in our patient. They were multiple and disseminated and their unusual appearance on chest radiographs, consistent with aggressive disease, initially imposed a DD with diffuse pneumopathy.

Nephroblastoma is by far the most obvious clinical hypothesis in a young child with tumor of the upper abdomen and lung metastases. The Wilms' tumor is the second most widespread abdominal tumor in childhood with up to 30% of cases as stage IV at diagnosis and lungs as the most common metastatic site. Although recent SIOP nephroblastoma protocols do not require histologic confirmation before the start of preoperative chemotherapy, the specific radiologic appearance and the ruling out of neuroblastoma on the basis of normal catecholamine urinary excretion are mandatory for diagnosis. A careful examination of the abdominal CT scan and a measurement of urine catecholamines would have differentiated neuroblastoma from Wilms' tumor in our patient at the initial evaluation.

The low incidence of lung metastasis in neuroblastoma is somewhat unique, as the lung is the most common site of metastasis in most other pediatric solid tumors. Patients presenting with lung dissemination at diagnosis have tumors with unfavorable histology, N-myc oncogene amplification and higher LDH levels. The event-free and overall survival are lower in these patients as compared to all other children with stage IV disease.

Conclusion

Pulmonary metastases at diagnosis are uncommon in stage IV neuroblastoma and reflect a biologically aggressive disease. We emphasize that abdominal neuroblastoma with lung dissemination in young children may be confused with stage IV Wilms' tumor.

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