

Intrarenal Neuroblastoma: A Diagnostic Challenge

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ABSTRACT

Abdominal masses in children are usually retroperitoneal in location and 50% are renal in origin. In infants less than 2 months, majority of renal masses are benign; frequency of malignancy increases with age. Wilms' tumor is the commonest solid intrarenal tumors. However, occasionally neuroblastoma occurs in the kidney. Neuroblastoma in 50% of cases presents before 2 years of age, and 75% before 4 years. Neuroblastomas can occur anywhere along the neuroectodermal sympathetic chain, although adrenal medulla is the commonest site (50 - 70%). Intrarenal neuroblastoma is a rare clinical diagnosis. It clinically and radiologically mimics Wilms' tumor and it is difficult to differentiate between the two pre-operatively. Lung metastasis and vascular invasion are also rare in neuroblastoma but rather common in Wilms' tumor. We present case of a patient who had extensive renal involvement with neuroblastoma and pulmonary metastasis, clinically and radiologically it was difficult to differentiate between the Wilms' tumor and neuroblastoma.

Key Words: *Intra renal. Neuroblastoma. Pulmonary metastasis. Abdominal mass.*

INTRODUCTION

Neuroblastoma and Wilm's tumor are two close differentials in children with renal mass. Neuroblastomas typically presents at 2 years with metastasis in 50% at presentation. Wilms' tumor presents between 4 - 5 years. Metastasis is usually to the lungs or inferior vena cava (IVC).¹ Intrarenal neuroblastoma is very rare and originates from either adrenal rests present within the renal tissue or from intrarenal sympathetic ganglia.² Differentiating intrarenal neuroblastoma from wilms tumor with pulmonary metastasis can create significant diagnostic challenge.

Hereby, we present the case of a 1.5-year boy who presented with a lumbar mass and created much diagnostic challenge.

CASE REPORT

A 1.5-year malnourished male toddler presented to the haem/oncology ward with 15-day history of left flank mass. On examination, the growth parameters were below the 5th centile for age. He was hypertensive, having blood pressure of 125/85. Abdominal examination revealed a 9 by 7 cm mass palpable in the left lumbar region. Imaging studies showed it to be a left renal mass with thrombus in renal vein and IVC reaching right atrium.

The clinical diagnosis of Wilms' tumor was made and pre-operative chemotherapy was started according to UKCCSG protocol.

Ultrasound guided trucut biopsy was done; histopathology showed it to be a round blue cell tumor. The 24-hour catecholamine levels were not determined due to prior chemotherapy which he had already received.

Since clinical and radiological diagnosis was not consistent with the histopathological diagnosis, so immunochemistry was sent that turned out to be NSE positive and WT-1 negative, which confirmed the diagnosis of intrarenal neuroblastoma with IVC thrombus. The parents were counselled and further workup was planned for intrarenal neuroblastoma. Unfortunately, the child became septic with broncho-pneumonia and expired.

DISCUSSION

Neuroblastoma, the most common extracranial solid tumor in children, is an embryonal malignancy of the sympathetic nervous system that arises from neuroblasts (pluripotent sympathetic cells). The most common site is suprarenal. Intrarenal origin with pulmonary metastasis is very rare presentation. Involvement of inferior vena cava by neuroblastoma is even rarer.

Intrarenal neuroblastoma is a rare and aggressive malignancy. Amplification of the N-MYC oncogene, the best-characterized molecular aberration, is present in 20% of cases and is a negative prognostic marker. Anaplastic lymphoma kinase (ALK) has also been implicated in neuroblastoma. Activating mutations in ALK have been reported in both familial and sporadic neuroblastoma.² Only 6 cases in adults and 8 cases in children have been reported in published studies.³ It is believed to originate from either sequestration of adrenal medullary tissue in the kidney during the fetal

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Received: September 12, 2013; Accepted: October 18, 2014.

development or from intrarenal sympathetic ganglion or results from direct renal invasion from an adrenal neuroblastoma. Patients are often hypertensive. It has poor long term survival.⁴ In one series, intrarenal neuroblastoma represented 3.8% of all non-Wilms' patients.⁵

Metastases are common at the time of presentation in neuroblastoma and may be the reason for presentation in up to half the cases. Typical sites of metastasis include bone, regional lymph nodes, orbit, and liver; but not lung.⁶ Lung metastases are rarely detected in children with neuroblastoma, seen in only 3.6% of 2,808 patients with Stage IV neuroblastoma patients in one large series.⁷ Lung metastases are more common among neuroblastoma patients with N-MYC amplified tumors, adrenal primary tumors, elevated LDH levels, and concomitant metastases to the CNS or liver.⁸

Vascular invasion is very rare in neuroblastoma, but common in Wilms' tumor. Neuroblastoma typically encases and displaces the vessels. Only a few cases have been reported in the literature showing tumor thrombus in IVC and right atrium by neuroblastoma.⁹

It is difficult to differentiate clinically and radiologically between intrarenal neuroblastoma and Wilms' tumor especially in index case where lung metastasis and IVC thrombus was also present. Diagnosis is usually based on imaging studies, urinary catecholamine, histopathology and immunochemistry. In the index patient clinical and radiological features were in favour of Wilms' tumor but histopathology was NSE positive favoring neuroblastoma. The 24-hour urinary catecholamine and MYCN-amplification studies was not possible due to resource constraint.

Similar to the prognosis of metastatic neuroblastoma, intrarenal neuroblastoma is a very aggressive tumor and can also present with bone metastasis.¹⁰ Treatment involves multiagent chemotherapy, surgery and irradiation with stem cell rescue.

Intrarenal neuroblastoma with pulmonary metastasis and IVC extension is a rare diagnosis. It can easily be

confused with stage IV Wilms' tumor especially in a setup with limited available diagnostic facilities. A strong clinical suspicion is required to differentiate between intrarenal neuroblastoma and Wilms' tumor, as prognosis and treatment of both these entities is entirely different. Imaging studies like ultrasonography and CT scan are often used in the initial evaluation of an abdominal mass so radiologists must also be aware of the appearance of this unusual manifestation of neuroblastoma on imaging studies.

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