Renal cell carcinoma in young girl with distant metastasis: a case report

This is a case of a 13-year-old female patient with flank pain and abdominal distention.

Imaging Findings:

Contiguous axial tomographic sections of the whole abdomen with intravenous contrast shows a large complex, inhomogeneously enhancing mass, measuring 17.4 x 13.9 x 13.4 cm (L x W x AP) arising from the middle to superior aspect of the right kidney. The mass has minimal central calcifications and central necrosis. It displaces the right liver lobe and adrenal superiority, bowels anteriorly and to the left side. The IVC and right renal vein are displaced leftwards with filling defects.

There are also multiple hypoenhancing lesions in the liver, the largest measuring 2.2 x 2.0 cm, likely representing liver metastases. There are also nodules with various sizes in the included lungs with the largest in the lateral segment of the right middle lobe measuring 2.2 x 1.8 cm, likely representing lung metastasis.

Biopsy done and sent for immunohistochemical staining for cytokeratin shows positive staining in 90% of tumour cells with strong reactivity, findings consistent with renal cell carcinoma.

Discussion:

Renal cell carcinoma is common in adult and represents 2% of malignant tumours in adults [2]. RCC is extremely rare malignancy in children and is estimated as 0.1% to 0.3% of all neoplasms and 1.8% to 6.3% of all malignant renal tumours, while the incidence of Wilms' tumour is 58% to 87% [3]. Studies shows that RCC accounted for 1.4% of all renal tumours in patients younger than 4 years, 15.2% in patients aged between 5 to 9 years, and 52.6% in patients aged 10 to 15 years [4].

There is no sex predominance for renal cell carcinoma in children but male predominance in adults. It is difficult to differentiate between RCC and Wilms' tumour preoperatively. Wilms' tumour is more common in children younger than 5 years of age and mostly presenting around 3 years of age, whereas RCC presents around the age of 8 to 9 years. Tumour calcification on plain X-ray or CT is more common in RCC approximately 25% than in Wilms' tumour 5% [3].

The most common symptoms are flank or abdominal pain, gross haematuria, and an abdominal mass. Other less
frequent symptoms are anaemia, fever, nausea/vomiting, pallor, malaise and weight loss [3].

RCCs comprise a heterogeneous group of tumours. The most common are the Xp11 (TFE3) translocation-RCC (20–40% of total), followed by papillary RCC, which comprises approximately 30% of total and includes both type 1 and type 2 lesions [1]. The diagnosis of paediatric RCC is made by considering pertinent clinical features, imaging findings and the histological appearance, supplemented by immunohistochemistry.

Incidence of metastases is similar to adults with half of the patients presenting with metastatic disease at the time of diagnosis. Radical nephrectomy is the treatment of choice for any stage. Post-operatively, radiotherapy and chemotherapy have been used in patients with advanced stage. The overall prognosis in children with RCC appears to be similar to adults and the most important prognostic factor is tumour staging. Five-year survival rate for patients with stage I is 92.4%, 84.6% stage II, 72.7% stage III and only 13.9% for patients with stage IV [2].

Although RCC is rare in children, clinical suspicion of this disease in children older than 5 years of age with renal masses is very important since the diagnostic and therapeutic approach differs from that for Wilms’ tumour. In our case CT findings of tumour with positive staining in 90% of tumour cells with cytokeratin was strongly suggestive of renal cell carcinoma.

**Differential Diagnosis List:** Renal cell carcinoma with liver and lung metastases, Wilms’ tumour, Pheochromocytoma

**Final Diagnosis:** Renal cell carcinoma with liver and lung metastases

**References:**

Description: Coronal plain reformatted image of the abdomen shows a large right renal mass with foci of calcification and central areas of hypointensity representing necrosis. Origin: DOI manila med medical center manila
b Description: Axial post-contrast image of the abdomen shows a large heterogeneously enhancing mass with mass effect on adjacent structure. Origin: DOI manila med medical center manila

c Description: Axial portal venous phase CT image of the abdomen shows a large heterogeneously enhancing mass displacing the IVC and right renal vein anteriorly with filling defect. Origin: DOI manila med medical center manila
Figure 2

**Description:** Axial portal venous phase CT of the abdomen shows multiple hypoenhancing lesions in the right hepatic lobe, likely representing liver metastases. **Origin:** DOI manila med medical center manila
**Description:** Axial CT of the included chest with lung window shows multiple nodules, likely representing lung metastases. **Origin:** DOI manila med medical center manila