BILATERAL NEPHROBLASTOMATOSIS: A ROLE FOR MULTISLICE CT

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ABSTRACT

Nephroblastomatosis is an abnormality of nephrogenesis characterized by incomplete maturation of the primitive nephrogenic cells. We report on one case of bilateral nephroblastomatosis in an 11-month-old boy who had an abdominal mass on the left side for 8 months. The boy had no other physical anomalies; he appeared healthy and had good development. A sonogram and an intravenous urogram revealed diffuse enlargement of both kidneys and ill-defined hyperechoic lesions covering both kidneys, except the lower pole of the right kidney. A multislice CT scan, performed with and without contrast medium, took no more than 10 minutes. We observed extensive enlargement of both kidneys and multiple solid masses throughout the left kidney and the upper half of the right kidney. Reconstruction on multi-planar views of both renal masses gave as good or better than MRI for displaying the extension of renal masses and renal vascular status. Surgical and pathologic findings are discussed in more detail.

INTRODUCTION

Nephroblastomatosis (NB) is an abnormality of nephrogenesis characterized by incomplete maturation of the primitive nephrogenic cells. The term was coined by HOU and Holman in 1961.[1] It also defines persistent metanephric blastema from infancy into childhood. In 1990, Bechwick et al. suggested that all foci of persistent metanephric tissue be referred to as nephrogenic rests and that the presence of a multiple nephrogenic rest be termed nephroblastomatosis.[2] The association of independent foci of persistent blastema with Wilms' tumor has been documented often. Minor degrees of NB are common with unilateral Wilms' tumour and are almost universal in bilateral Wilms' tumour. The most severe cases have features of both developmental malformation and neoplasia. Therefore, NB is widely accepted as a premalignant lesion.[3] The role of imaging in evaluating patients suspected of having NB, Wilms' tumour, or both, includes preoperative assessment of both kidneys, follow-up of the patient with known micro- or macroscopic NB to detect neoplastic changes, and screening of patients with syndromes associated with NB and Wilms' tumour. Microscopic foci of NB are not identified with any imaging modality.[3] In the case of macroscopic NB, diagnosis by ultrasound and excretory urography is less sensitive than computed tomography (CT) and magnetic resonance imaging (MRI). The use of spiral CT has definitely improved the efficacy of CT because of the more rapid scanning time, multi-planar reconstruction, and lower radiation doses particularly when multislice CT is performed.[4] We report one case of bilateral NB examined by multislice CT. We discuss the advantages of this technique in both diagnosing and preoperative assessment of tumor extension and its vascular supply of both kidneys.

CASE REPORT

An 11-month-old boy came to hospital with a left upper quadrant mass present since he

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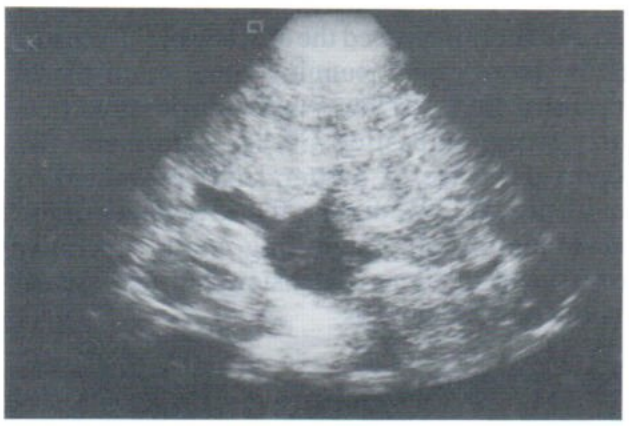
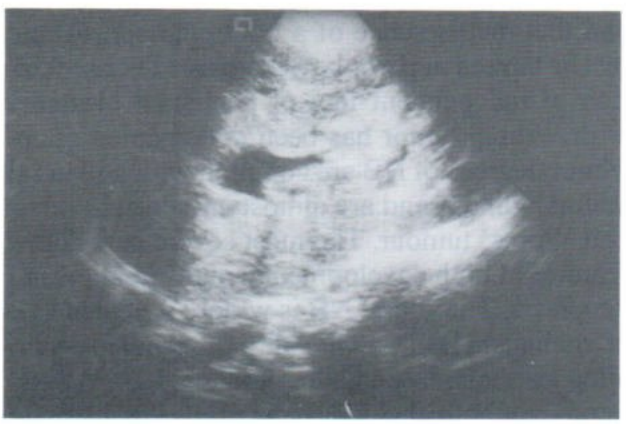
was 2-month-old. He had hyperpigmented skin and nevus of his back and neck, respectively. Other physical findings, laboratory tests, past medical and perinatal history were normal. There were no accompanying congenital abnormalites. The ultrasonogram revealed enlarged kidneys bilaterally with heterogeneous hyperechoic lesions covering the entire left kidney and the upper half of the right one (Fig.1a,b). Intravenous urography also revealed enlargement of both kidneys with compressed and distorted calices (Fig.2a,b). These findings were initially interpreted as polycystic kidney disease. At the follow-up three months later, the left kidney still had a hard consistency and irregular surface, so we performed a further multislice CT.

Sedation using oral chloral hydrate (0.5 mg/kg) was administrated before scanning. For the precontrast scan, the multislice CT (SOMATOM Plus 4 Volume Zoom, Siemens) was performed with 5-mm wide slices, 2.5-mm thick collimation, 12.5 mm/s table speed (pitch = 6) and 5 mm reconstruction interval. After intravenous administration of a nonionic contrast medium (2 mL/kg) performed using a mechanical injector at 2 mL/s, the volume scan began at the end of the mechanical injection and 70 seconds after the injection. The multislice CT scan was performed with 1.25-mm wide slices, 1-mm thick collimation, 5mm/s table speed (pitch = 5), and 5-mm reconstruction intervals. Fifty-five mAs and 120KV were performed for all three scans. The total scan time, including contrast medium injection, was less than 10 minutes. Multi planar reconstruction (MPR) and maximun intensity projection (MIP) were performed after finishing the axial image reconstruction. The CT of the abdomen revealed multiple nonenhancing cortical solid masses, an enlarged, surficially nodulated left kidney and upper half of the right (Fig.3). MIP and MPR clearly revealed the main renal arteries and veins and their branches into the renal masses (Fig.4). These images also showed the extension of the NB involving the entire left kidney and the upper half of the right kidney. A left nephrectomy was performed and revealed a10-cm, well encapsulated, nodular surficial mass covering the left kidney (Fig.5a,b). Cortical lobulated masses occupied the upper half of the right kidney and were biopsied. The histopathology was nephroblastomatosis (NB) of the left kidney with foci of Wilms' tumour and NB of a piece of the right kidney (Fig.6). Additional chemotherapy with vincristine and actinomycin was used to treat the stage I Wilms' tumour. The patient was referred to their nearest hospital for follow-up of the right kidney in case of tumour recurrence on left renal bed.

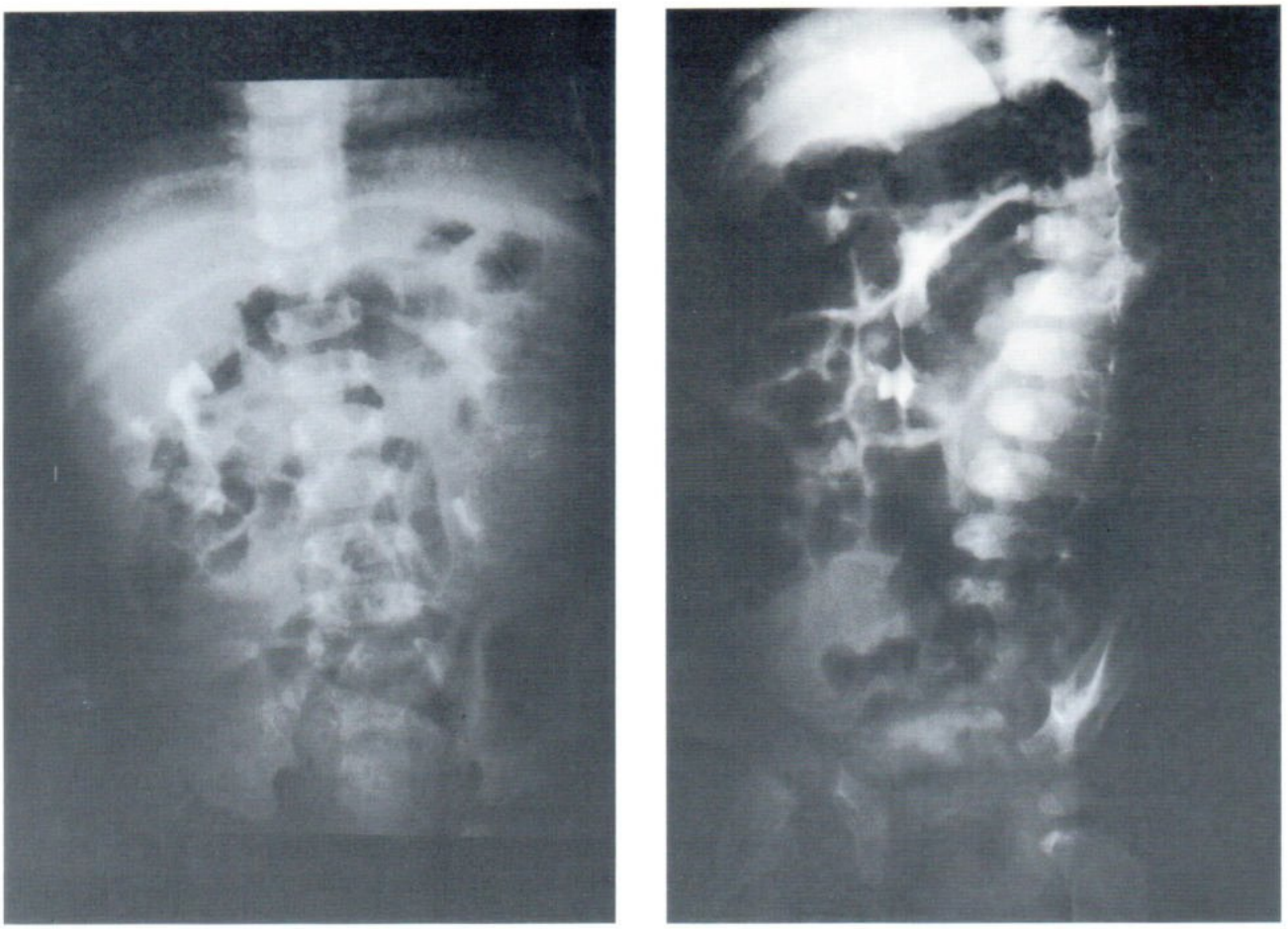


1A



1B

Fig. 1 a,b. Sonogram reveals enlarged kidneys bilaterally with heterogeneously hyperechogenic lesions and some degree of caliectasis on the entire left kidney (a) and a mass at the upper pole of the right kidney (b).



2A 2B

Fig. 2 a,b. Intravenous urography reveals an enlargement of both kidneys with compressed and distorted calices on the anteroposterior (a) and right posterior obligque (b) views.

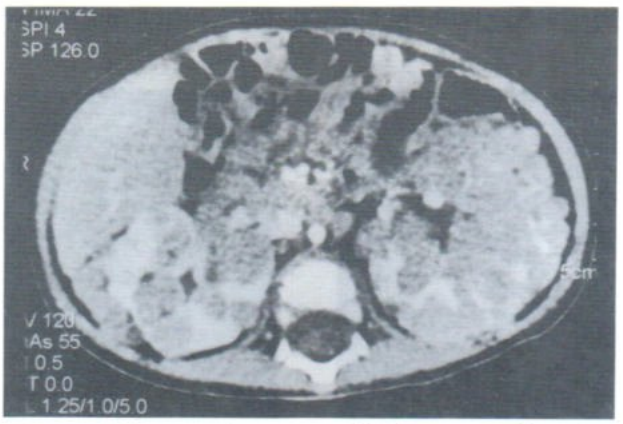
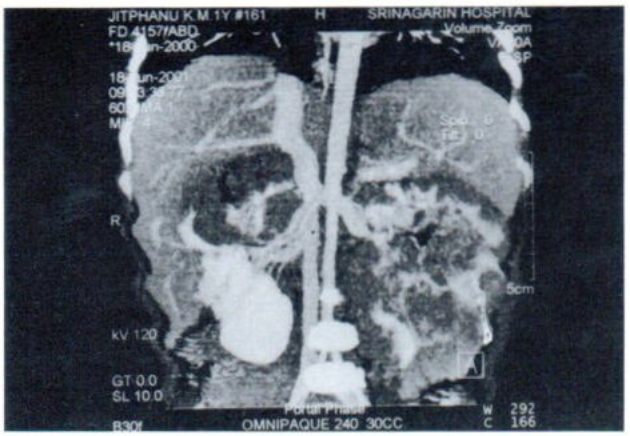
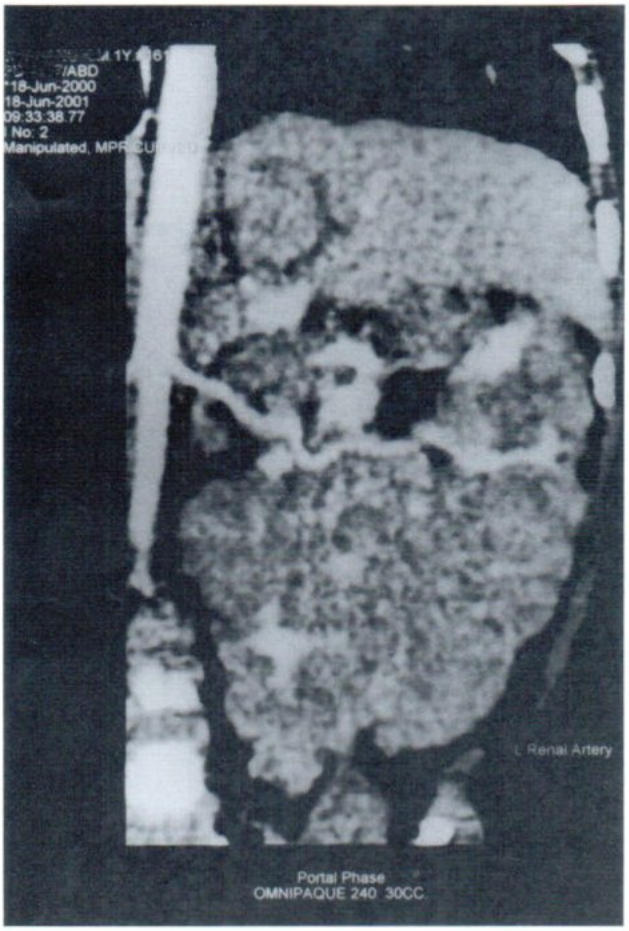


Fig. 3. CT scan shows multiple non-enhancing masses in the renal cortex of both kidneys, larger and more confluence in the left kidney with compressing and distorting of the collecting system.

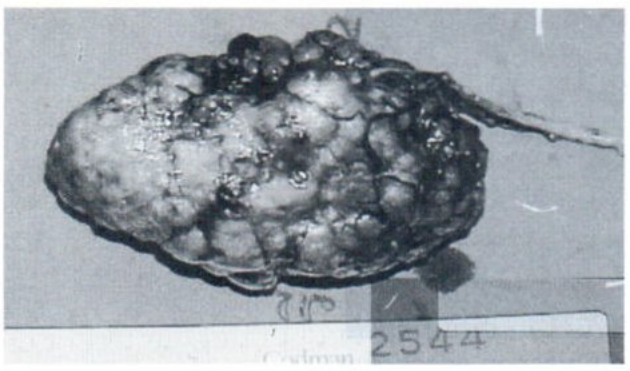


4A

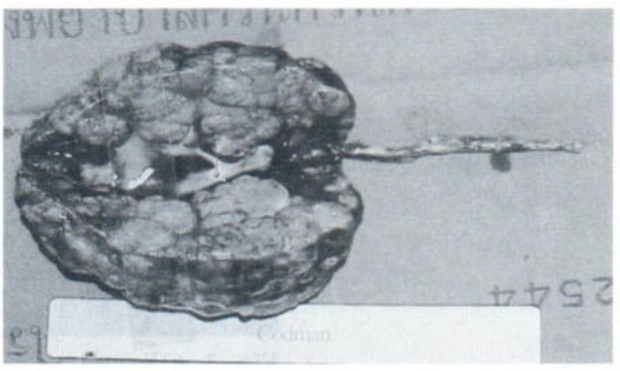


4B

Fig. 4 a,b. Maximum Intensity Projection (MIP) gives an excellent rendition of both renal arteries and veins (a). Multi-planar Reconstruction (MPR) reveals a branch of the left renal artery within NB of the left kidney (b). Both MIP and MPR also provide an extension of NB in both kidneys and the normal renal cortex at the lower pole of the right kidney.



5A



5B

Fig. 5 a,b. A left nephrectomy revealed a 10-cm well encapsulated, nodulated surface mass (a) while the cut surface revealed multiple masses throughout the entire renal cortex (b).

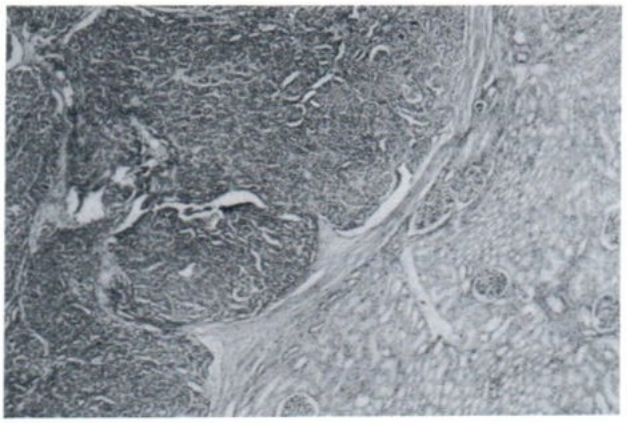


Fig. 6. Lower power photomicrograph shows multiple perilobar NB of variable sizes and well delineates from the normal renal parenchyma (right hand side).

DISCUSSION

The presence of multiple or diffuse nephrogenic rests define nephro-blastomatosis (NB). If nephrogenic rests persist until birth and during infancy, they usually form a Wilms' tumour so are regarded as precursor lesions.[3] Bechwith et al. suggested a modification in the classification of lesions with respect to their distribution within the renal lobe and classified NB as perilobar, intralobar and diffuse. The number of nephrogenic rests are classed: unifocal, multifocal, or diffuse.[2] In an attempt to divide lesions into clinically relevant groupings, the individual lesions are further subclassified as dormant, maturing/sclerosing, hyperplastic, and neoplastic. This subclassification requires both microscopic and gross pathologic evaluation of the lesions.[3]

Imaging findings of nephroblastomatosis (NB) typically reveal homogenously isoechogenic or slightly hypoechogenic nodules compared to the renal cortex[6,7,8] but unlike our case in whom that heterogeneously hyperechogenic lesions were demonstrated. The excretory urogram and CT findings of NB in our patient were similar to other reports.[3,4] Rohrschneider et al. reported 12 cases of NB checked with MRI, which revealed isointense or slightly hypointense to the cortex on T1W, T2W and proton-density images with poor delineation. Gadolineum-enhanced images were best used to detect lesions that remained homogeneously hypointense compared with the brightly enhancing cortex. They concluded, MR was the method of choice in view of significant radiation from serial CT.[6] Today CT scanners have been improved, particularly the multislice CT (SOMATOM Plus 4 Volume Zoom), in that one rotation produces 4 slices, so selecting a greater pitch to speed up acquisition and shorten scan time reduces radiation exposure. The authors also used low mAs in order to reduce radiation doses. The multislice CT provides thin slice reconstruction of the kidneys, which is an advantage in small NB thus allowing making possible a clear distinction between non-enhanced NB and enhanced normal renal parenchyma.

ACKNOWLEDGMENTS

The authors thank Mr. Bryan Roderick Hamman for assistance with the English-language presentation of the manuscript.

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