

The computed tomography features of multifocal nephroblastomatosis

Multifocal nephroblastomatosis has multiple smooth foci in the periphery of the renal cortex. These lesions are lenticular or ovoid, usually 5-20 mm in size and sharply demarcated. The kidneys are large and their contour is lobulated, often with significant bulges. In contrast, diffuse nephroblastomatosis produces a thick uniform peripheral rind of abnormal tissue in an enlarged kidney with preservation of the renal shape and contour.

At computed tomography (CT), the density of the lesions resembles normal cortex. These lesions are poorly vascularised and after contrast administration are markedly hypodense when compared with normal renal cortex. The centrally located normal

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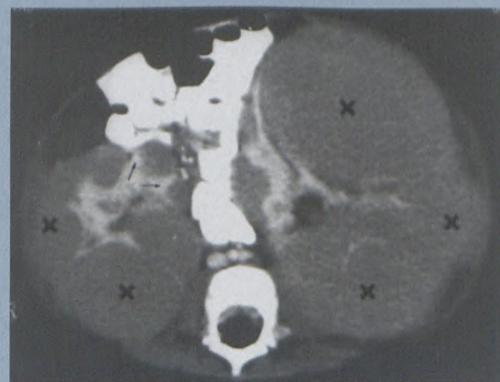
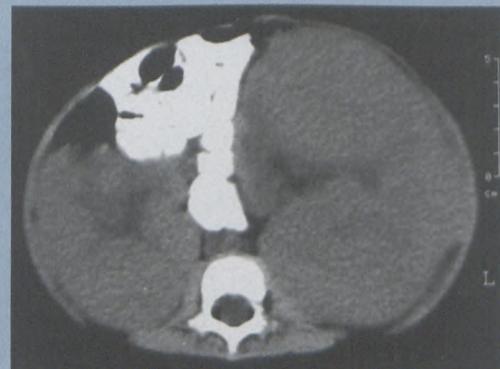
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Nephroblastomatosis refers to persistent metanephric blastema (nephrogenic) rests found within the kidneys after 34 weeks' gestation. Autopsy studies have shown that they occur in up to 1% of children under 3 months of age. They are considered premalignant owing to their strong association with Wilm's tumours. Even though unifocal and diffuse disease occurs, multifocal nephroblastomatosis is the most common type encountered. Lesions are located primarily at the periphery, but may occur anywhere within the kidney.

Effective management of these children requires identification of both the nephroblastomatosis and the Wilm's tumours. Imaging is critical, as surgical biopsy may be complicated by the multiplicity and small size of the lesions, as well as the risk of peritoneal seeding.



Figures 1a and 1b: Pre- and post-contrast CT of the kidneys. On the pre-contrast image, the left kidney is enlarged more than the right, but both demonstrate a homogeneous density. After contrast administration, the characteristic centrally located enhancing normal parenchyma resembling "stag antlers" (arrows) is well demonstrated against the non-enhancing peripheral multifocal nephrogenic rests (crosses).

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renal tissue shows characteristic distortion resembling "stag antlers" after contrast administration. The most characteristic feature of the nephrogenic rests is their homogeneous appearance. In comparison, Wilm's tumours are often heterogeneous both pre- and post-contrast administration. Size and shape are unhelpful features in differentiation. Lymphoma may produce homogeneously enlarged kidneys that may initially appear identical to diffuse or multifocal nephroblastomatosis, but will not demonstrate the central "stag antlers".

CT is considered to have similar potential to gadolinium-enhanced MRI in the diagnosis of nephroblastomatosis with regard to identification and differentiation from Wilm's tumour. In our personal experience, however, MRI has a greater sensitivity for the detection of small lesions.

References

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Figures 2a and 2b: Before and after chemotherapy. Typical homogeneous peripheral nephrogenic rests with central "stag antler" configuration of the contrast-enhancing normal parenchyma. The post-chemotherapy CT scan shows a reduction in the size of the nephroblastomatosis, with expansion of the previously compressed normal parenchyma.