

Ossifying renal tumor of infancy

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ABSTRACT

Ossifying renal tumor of infancy (ORTI) is a benign neoplasm, which presents with gross hematuria and less frequently as an abdominal mass, histologically it comprises a large cell with an epithelial nature and osteoid formation. We report a case of a 10-month-old girl who developed ORTI as non-opacified upper calyces of left kidney on excretory urography. We outlined the calcific and tumors nature of the lesion by ultrasound and computed tomography. The macroscopic and histological features were pathognomonic.

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Ossifying renal tumor of infancy (ORTI) is a rare but distinctive tumor that presents frequently as a mass in the pelvicaliceal system.¹ More than 11 cases are reported in the literature.² Lowe et al,³ differentiated this report from other pediatric masses by the presence of ossified elements.¹ Soletto-Avila et al,² thought the tumor to be a distinctive clinico pathologic entity from interaction between hyperplastic intralobar nephrogenic rests (ILNR) in the renal papilla with the urothelial cells in the distal collecting ducts,² considered biologically benign.^{1,4} Chitten et al,⁵ first described it in 1980. We present a rare case of ORTI in an infant. We describe the imaging features however, confirmed the diagnosis only by histopathologic examination.

Case Report. A 10-month-old girl was admitted with bronchial asthma, she had 2 episodes of hematuria while in the hospital. Physical examination was unremarkable. Urine microscopy, other than hematuria was normal, serum urea, creatinine, calcium, and phosphate were within normal limits. There was no growth on urine culture. Ultrasound (US) revealed a hyperechoic mass of 18 x 13 mm in the upper pole of the left kidney with distal acoustic shadow suggesting

possible calculus, however, excretory urography showed no calcification, and non visualized upper renal calyces, which raised the possibility of a tumor. Computed tomography (CT) on the other hand, confirmed the presence of an ossified lesion in the upper pole, and showed a radiolucent halo after IV contrast enhancement (**Figures 1a & 2b**). The right kidney was normal. A nuclear study; namely, technetium 99m dimercaptosuccinic acid (99mTc-labelled DMSA) showed a differential renal function of 44% for the left kidney, and 56% for the right kidney. On the basis of the clinical and imaging features, the lesion was thought to be malignant, left radical nephrectomy was performed and the postoperative course was uneventful. We found a firm white partially ossified tumor measuring 2.5 x 1.5 x 1.0 cm with some features that mimicked a calculus in the upper pole of the left kidney; it arose from the renal medulla and was an integral part of the renal parenchyma (**Figure 2a**). Microscopically the lesion consisted of an amorphous mass of partially mineralized osteoid, primitive collecting duct-like tubules and plump polygonal cells, which were integral parts of the lesion (**Figure 2b**). A thin rim of immature cytologically bland spindle cells and primitive

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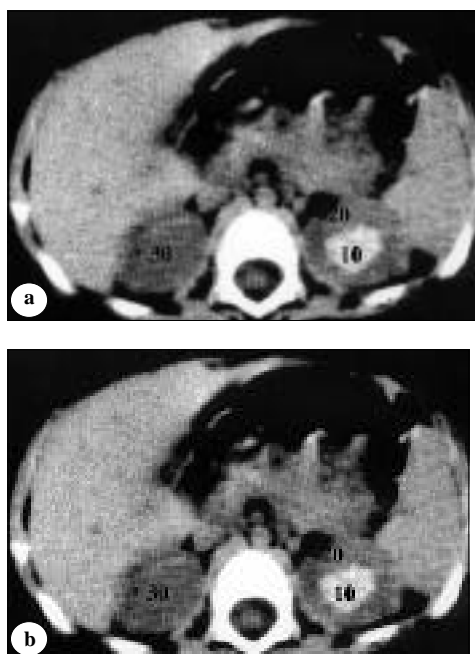


Figure 1 - CT scan of renal area: (a) Showing a well circumscribed calcified mass in the upper pole of the left kidney. (b) Enhanced CT demonstrate a non-enhancing radiolucent halo around the calcified upper renal mass.

tubules surrounded the central mass of osteoid. There were localized chronic pyelonephritic changes in the adjacent renal cortex. The attached lymph nodes showed non-specific reactive changes.

DISCUSSION. Gross hematuria is a major clinical feature of ORTI,² which may also present as a palpable abdominal mass.⁶ The radiological investigations play a vital role in the initial diagnosis. All lesions were attached to a renal papilla and presented mainly within the calyceal lumen or as a small nodular tumor protruded into the pelvis from the superior calyceal region.^{2,4} Some clinically resemble staghorn stone causing severe hydronephroses.^{2,6} All tumors contained a varying proportion of osteoid, osteoblastic cells and spindle cells. The proportion of osteoid and the degree of osseous maturation increases with increasing age of patients.² In our patient, US initially suggested the presence of foci with minimally calcified osteoid, while excretory urography showed a non visualized upper calyces, CT revealed its overall calcific nature explicitly, it is known to be more accurate than plain radiography in detecting the presence of calcium, moreover, CT scan showed a radio-lucent halo after intravenous contrast enhancement. This supported the concept of a partially ossified tumor as well. Imaging helps in the differential diagnosis for a patient of this age, which included Wilm's tumor, extra-adrenal neuroblastoma, infection, calculus, and

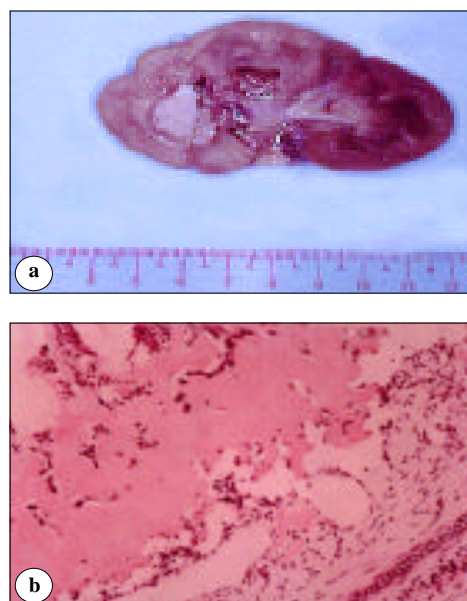


Figure 2 - Macroscopic pathology specimen of left kidney (a) showing a firm ovoid calcified tumor from the renal medulla with evident surrounding hydronephrosis. Histological specimen (b) showing an amorphous mass of poorly mineralized osteoid, plumps of polygonal tumor cells, spindle cells and immature tubules all as part of the lesion.

calcified hematoma.⁵ Lowe et al,³ differentiated it from mesoblastic nephroma by the presence of ossified elements. The macroscopic and histological features are pathognomonic.² The cases with adequate follow-up indicate that it is a benign tumor,^{2,4} the lesion probably arises from ILNR,² it can be multifocal and a precursor for nephroblastoma.^{2,3} We therefore recommended careful follow-up and surveillance of the contralateral kidney.

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