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Unusual Ossifying Pediatric Renal Mass: A Case Report

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Abstract
Renal cell carcinoma is an uncommon tumor in pediatric population.It is important for the clinicians to know its diagnosis as the management of renal cell carcinoma RCC is different from Wilms Tumor which is the most common tumor encountered in children .The management is surgical resection with no proven role for chemo or radiotherapy so far. Here, we present a case of seven-year-old male child presented with haematuria and no palpable lump. An echogenic mass was detected on his ultrasound. Further characterisation of the mass was studied by contrast enhanced ultrasound (CEUS), non-contrast computed tomography (CT), and magnetic resonance imaging (MRI). Final diagnosis was confirmed based on histopathological examination. Left nephroureterectomy was done.
Keywords: Renal cell Carcinoma, Wims tumor, Contrast Enhanced Ultrasound, Ossifying

Introduction

Renal masses in children have different histopathological identities. While Wilms's tumor is known to be common in first decade of human life, renal cell cancer (RCC) is not uncommon in the second decade onwards. RCC is rare in the first decade and is reported to constitutes 2 to 5% of all the malignant renal tumors in children (1). Various factors determine its prognosis, including age, histological subtype, staging, and the symptoms at presentation. Most common imaging presentation is solid renal mass; however, the imaging features might vary. Surgery is the mainstay of treatment for RCC. Here, we described an unusual imaging appearance of RCC which led to an imaging dilemma.

Case Report

A Seven-year-old male child presented to Pediatric Surgery Outpatient Department (OPD) with painless hematuria in the last 3 months. There was no history of abdominal trauma. On examination, abdomen was soft and no lump was palpable. Due to having a history of hematuria, imaging evaluation was started. Ultrasound was done which revealed well marginated echogenic lesion of size 3.2 x1.6 cm in the lower pole extending into the lower calyx and the renal pelvis with no significant color flow on colour Doppler (Figure 1a and b). Contrast enhanced ultrasound (CEUS) was done on Aixplorer (Supersonic Imagine, France) using ultrasound contrast agent (Sonovue, Bracco, Milan, Italy). Then, 2.4 ml of contrast was injected through the antecubital vein followed by 5ml of bolus saline injection. CEUS revealed that the mass to be enhancing in early phase and wash out in delayed phase (Figure 2a and b). The echogenic appearance of the mass led to diagnostic confusion. For further characterization, MRI was performed on a 1.5 T scanner (Acheiva 1.5 T, Philips, Netherlands). The mass was T1W isointense to cortex and showed no signal drop on opposed phase images. On T2W, the lesion was hypointense to cortex. Peripheral hypointense rim was seen on both T1 and T2W [Fig 3a and 3b] which

showed blooming on GRE images. This led to the suggestion of peripheral calcification or haemorrhage. The mass was showing mild restriction on DW images. In order to resolve the issues of calcification, limited NCCT was performed on a multidetector row CT scanner (Somatom Sensation 40, Siemens, Erlangen, Germany). An amorphous calcification of the mass and peripheral rim of calcification were seen on NCCT (Figures 4a and 4b). The mass was limited to kidney with no evidence of vascular invasion and no evidence of retroperitoneal lymphadenopathy. Right kidney was normal with no evidence of any focal lesion. No evidence of distant metastasis was seen. Ultrasound Guided Biopsy was done which revealed tumor with cells having abundant cytoplasm with moderate atypia and extensive psammomatous calcification. Tumor cells are immunopositive for vimentin and CD 10 negative for HMB 45 and Melan. All these features were consistent with Xp 11.2 translocation associated RCC (Figure 5). Hence, chromosomal studies were advised. Patient underwent left ureteronephrectomy. Post operative period was uneventful. The child came for follow up in OPD and did not have any complaints, so imaging follow-up was not done.

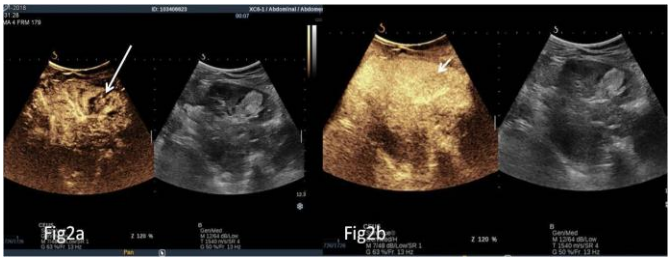

Figure 2(a and b). Contrast enhanced ultrasound shows the mass lesion has mild contrast enhancement relatively less than renal parenchyma with wash out.

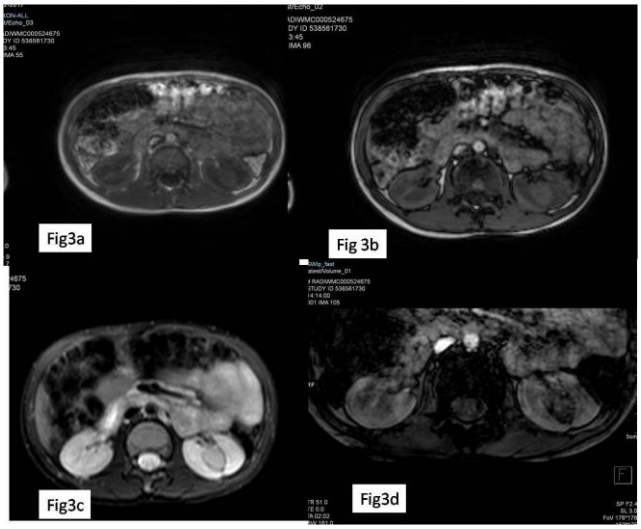

Figure 3(a-d) Axial T1W image shows that the mass lesion is isointense to renal parenchyma on T1W with no suppression on opposed phase image and hypointense on T2W with peripheral hypointense rim (arrow), and it also shows blooming on GRE.

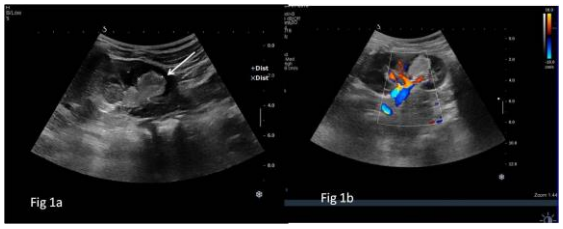

Figure (1a and b). Grey scale and color Doppler ultrasound depicting echogenic mass lesion (arrow) in the lower pole of left kidney. Minimal vascularity is seen on color flow.

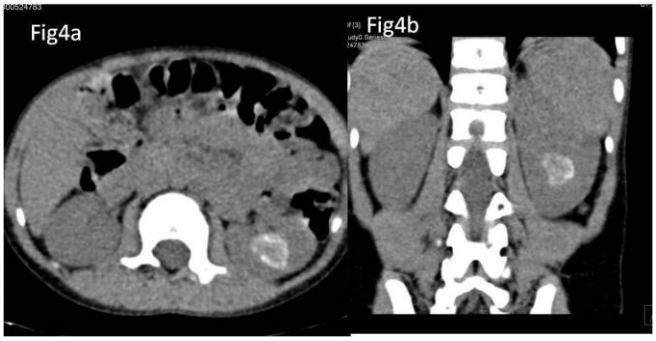

Figure 4(a and b) axial and coronal reformatted Non-contrast CT show that the mass has peripheral rim of calcification.

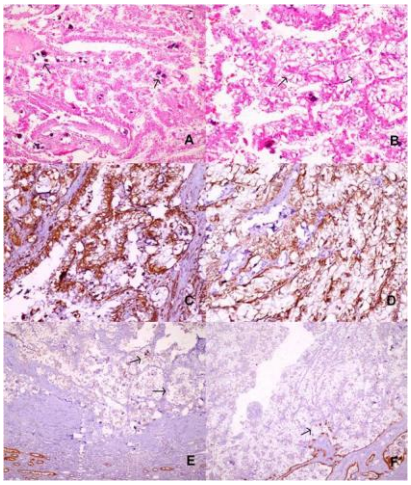

Figure 5. Photomicrographs show a renal tumour with papillary and alveolar pattern along with posamommatous calcific bodies (arrows) [A x 40]. The tumour comprises of polygonal tumour cells with voluminous clear to eosinophilic cytoplasm (arrows) [B x 200]. The tumour cells are strongly positive for vimentin [C x 200] and α-methyacryl coenzyme A racemase [D x 200] stains. The tumour cells are focally positive for CD10 stain (arrows). Strongly positive proximal renal tubules are noted in periphery of this figure [E x40]. Focal positivity for cytokeratin 7 stain is also noted (arrow) [F x 40]

Discusssion:

Renal masses constitute one of the commonest pediatric abdominal masses. Among all the pediatric renal tumors, Wilma's tumor is the most common which accounts for 90% of all the renal malignancies and 7% of the all the pediatric malignancies (1). Pediatric RCC is rare; however, the survival of pediatric RCC is 72% which is more than other non-Wilms renal tumour (NWRT) which is 63% (1). Peak age of Wilms tumour is 3 years old and RCC presents at the age of 8 to 9 years. Majority of the children affected with RCC have underlying genetic disorders like Von Hippel Lindau syndrome or have undergone chemotherapy or radiation for Wilms tumour or neuroblastoma (3). It is often incidentally detected through a mass or present with only hematuria or abdominal pain. RCCs are single and relatively small in size. However, bulky, multiple, bilateral, and metachronous lesions have also been reported (3). Imaging plays a significant role in determining the size, extent, vascular invasion, lymph node involvement, calcification, contralateral tumour, and metastasis. In this case, the mass was detected on abdominal ultrasound which was echogenic showing relatively less flow on color Doppler in comparison with the renal parenchyma. On imaging, in comparison with Wilms' tumour, RCCs are relatively smaller and have more frequency of peripheral calcification (4). In our patient, the type of calcification in NCCT led to the diagnostic confusion. This extensive calcification in a renal mass can be seen in ossifying renal tumor of infancy although the age group is different. Other imaging differential on ultrasound of an echogenic mass would be fat /haemorrhage containing lesion but these were ruled out by MRI and CT. On MRI, no specific feature has been described. In a case report by Hyung et al., the mass was low to intermediate signal intensity on T1W and intermediate to high signal on T2W (5). However, in our case, the mass was isointense on T1W and hypo intense on T2W with peripheral hypointense rim. T2W hypointense rim on MRI has not been described previously.

RCC originates from the epithelium of renal tubules. Histologically, RCC is divided into various subtypes papillary, clear cell, chromophobe, collecting duct, and unclassified type. Pediatric RCC is different from adult RCC as the translocation type is more common followed by papillary and medullary variety in contrast to adults where clear-cell cancer is more common followed by papillary. (6). One of the Xp translocated TEF 3 gene fusion associated with renal cell carcinoma is seen in the pediatric age group (7). According to WHO classification in 2004, Xp11.2 and TEF 3 fusion gene associated with RCC was classified as separate entity (8). Histologically, these tumours show voluminous papillary pattern with clear

cytoplasm, nested architecture, and granular eosinophilic cytoplasm. Hyaline nodules and psammoma bodies can be seen. So, Xp11.2 translocated RCC is confused with papillary RCC. Presentation in childhood, large tumour size, areas of cystic and necrotic changes, presence of calcification, high attenuation on NCCT, presence of lymphadenopathy, and distant metastasis favour Xp11.2 RCC (9).

On imaging, translocation RCC's appear heterogeneous with the presence of solid and cystic components with areas of hemorrhage, necrosis, and calcifications. Most of these lesions are located in the medulla. Exophytic component is rarely seen. On ultrasound, these tumours are heterogeneous with the presence of egg shell calcification. On MRI, the tumour is hypointense to cortexon T2W with heterogeneous due to fat, haemorrhage, and cystic component, and enhances heterogeneously but less than that of renal parenchyma (6). In our patient, the tumour was calcified and high attenuation on NCCT. On MRI, the lesion was hypointense on T2W and isointense on T1w with peripheral hypointense rim.

The prognosis of RCC in pediatric age group is better than other non-Wilms' renal tumour (NWRTs) and has the highest survival rate (1). Surgical resection is the only management in contrast to the management of Wilms' tumour where chemotherapy plays a significant role. The survival of these tumours post resection depends on the completeness of the resection which can be as low as 10% in case of incomplete resection. Majority of recurrences or death occur within 2 years; however, late recurrence has also been reported. The management differs from adult RCC in which nephron saving surgery can be done. Role of chemotherapy or immunotherapy has not been clear in this regard. Post-operative chemotherapy or radiotherapy has been used for the higher grade tumours.

Till date, very few case reports have been published on the imaging appearance of Xp associated with RCC in children on CEUS and MRI. Our case briefly described the imaging appearance on CEUS and MRI.

Conclusion

To conclude, RCC is a rare tumour in children; however, it should be considered when the child age is more than 5 years. The translocation variety can be suspected if high contrast attenuation on CT is noted. It is important as the prognosis of RCC in childhood is better than NWRT.

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Conflicts of Interest

No conflict of interest exits between Dr Poonam sherwani and Co-authors.

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