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Case Study

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UNCLASSIFIED TYPE OF RCC- A CASE REPORT

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INTRODUCTION

According to the WHO 2016 classification of renal tumors, unclassified RCC is a diagnostic category which does not fit any of the well recognized subtypes of RCC.[1] It is a rare subtype and represents 2–6% of renal epithelial tumors in adults.[2] It shares variable morphological features that overlap considerably with other subtypes. Imaging plays an important in diagnosing early stages of RCC, besides helping in staging and therapeutic planning.[3]

CASE REPORT

A 64 year old female presented with complaint of painless abdominal lump, progressively increasing in size since 2018. No history of hematuria. Laparotomy done in a peripheral centre. However, abdomen closed without taking any biopsy sample. Patient was later referred to our hospital. Patient was non compliant with the workup. She got her first CECT abdomen done in February 2021 and was planned for surgery, however patient was lost to follow up. Repeat CT was done in September 2021. There were no significant clinical complaints.

Contrast enhanced images reveal a large solid cystic mass in the abdominal cavity. It has predominantly cystic component with enhancing solid areas seen at the periphery and in the central part. The left kidney is displaced anterolaterally and seen to form claw with left kidney with compression of left renal vein with proximal distension. No intraluminal thrombus in left renal vein or artery seen.

On comparison with the previous scan done in September 2021, no significant interval growth/ vascular invasion/ infiltration into the adjacent structures seen. No lymphadenopathy or any metastasis seen. The final diagnosis based on imaging was made of left renal mass with central necrosis, without any vascular infiltration (s/o Renal cell carcinoma).

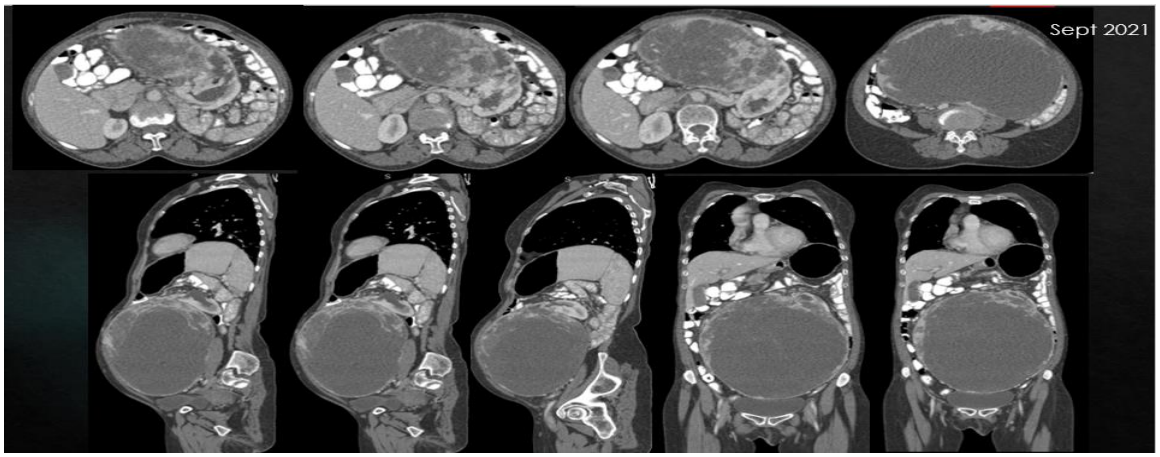


Figure 1(a)

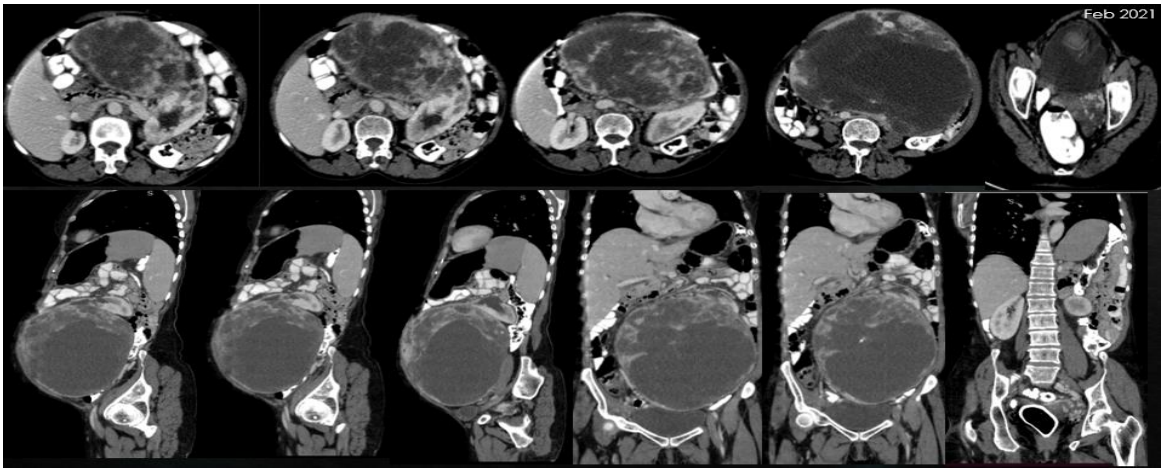
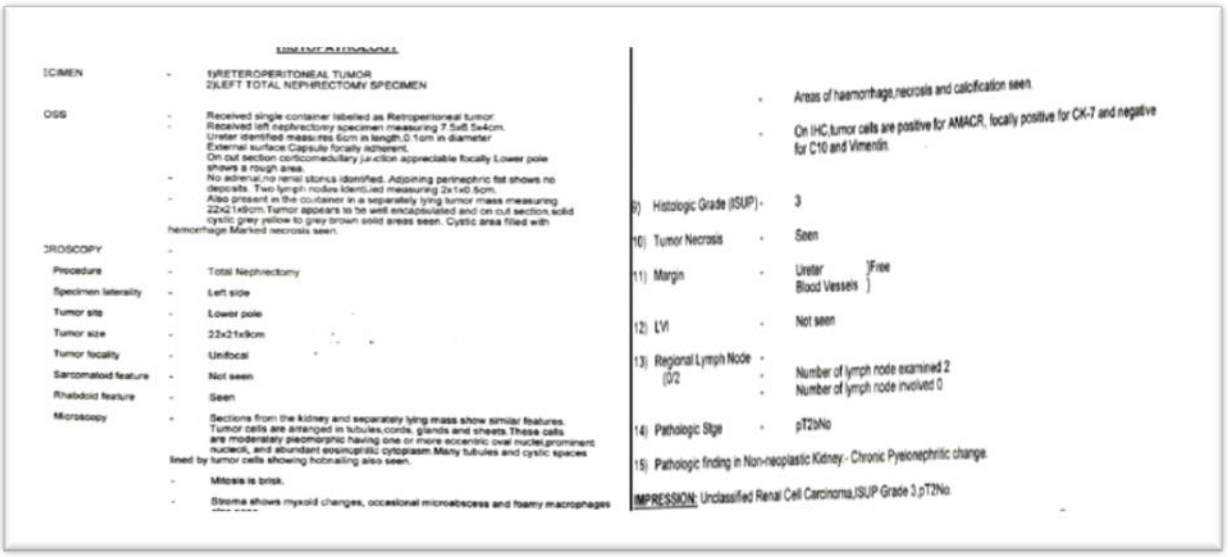


Figure 1(b)

Figure 1: CECT Abdomen in Figure 1(b) reveals a large solid cystic mass with predominant cystic component showing enhancing solid areas at the periphery and in the central part and is seen making claw with left kidney. Comparison of both the scans[Figure 1(a), Figure 1(b)] reveal no significant interval change.



ICIMEN - 1)RETEROPERITONEAL TUMOR
2)LEFT TOTAL NEPHRECTOMY SPECIMEN

OSS - Received single container labelled as Retroperitoneal tumor.
- Received left nephrectomy specimen measuring 7.5x6.5x4cm. Ureter identified measuring 6cm in length,0.1cm in diameter External surface Capsule focally ichherent. On cut section corticomedullary junction appreciable focally Lower pole shows a rough area.
- No adrenal,no renal stones identified. Adjoining perinephric fat shows no deposits. Two lymph nodes identified measuring 2x1x0.5cm.
- Also present in the container in a separately lying tumor mass measuring 22x21x9cm. Tumor appears to be well encapsulated and on cut section,solid cystic grey yellow to grey brown solid areas seen. Cystic area filled with hemorrhage.Marked necrosis seen.

ROSCOPY -

Procedure	-	Total Nephrectomy
Specimen laterality	-	Left side
Tumor site	-	Lower pole
Tumor size	-	22x21x9cm
Tumor focality	-	Unifocal
Sarcomatoid feature	-	Not seen
Rhabdoid feature	-	Seen
Microscopy	-	Sections from the kidney and separately lying mass show similar features. Tumor cells are arranged in tubules,cords, glands and sheets.These cells are moderately pleomorphic having one or more eccentric oval nuclei,prominent nucleoli, and abundant eosinophilic cytoplasm.Many tubules and cystic spaces lined by tumor cells showing hobnailing also seen.
	-	Mitosis is brisk.
	-	Stroma shows myxoid changes, occasional microabscess and foamy macrophages
	-	Areas of haemorrhage,necrosis and calcification seen.
	-	On IHC,tumor cells are positive for AMACR, locally positive for CK-7 and negative for C10 and Vimentin.

9) Histologic Grade (ISUP) - 3
10) Tumor Necrosis - Seen
11) Margin - Ureter, Blood Vessels } Free
12) LVI - Not seen
13) Regional Lymph Node (0/2 - Number of lymph node examined 2, Number of lymph node involved 0
14) Pathologic Stge - pT2bNo
15) Pathologic finding in Non-neoplastic Kidney - Chronic Pyelonephritic change.

IMPRESSION: Unclassified Renal Cell Carcinoma,ISUP Grade 3,pT2No.

Figure 2(a)

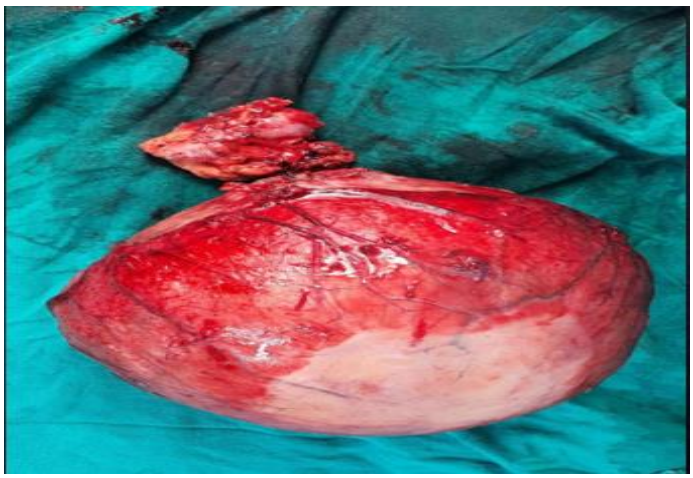


Figure 2(b)

Figure 2. Final Histopathological report suggestive of Unclassified Renal Cell Carcinoma, ISUP Grade 3 as shown in Figure 2(a). Surgical specimen of the mass lesion showsn in Figure 2(b).
Image courtesy: Department of surgical oncology, VMMC & SJH, Delhi.

DISCUSSION

Unclassified RCC(uRCCs) is not a distinct type of RCC but a heterogeneous group of tumours with variable clinicopathologic features and biological behavior. These comprise less than 5 % of all RCCs.[4] It is actually a histopathological diagnosis of exclusion. Usually has a poor outcome with no standard therapy (needs surgical resection only). uRCCs usually show high-grade histologic features.[4] Imaging methods can also differentiate in clear cell and non clear cell subtypes of RCC, mainly by evaluation of density, signal intensity and pattern of intravenous contrast uptake.[5] Unclassified RCC are increasingly being recognised due to morphologic overlap between clear cell, papillary and oncocytic subtype; and include both low grade and high grade histological subtypes.[6]

Metastatic potential is higher in these tumours and sarcomatoid changes suggests poorer prognosis.[7] As compared to clear cell variety, unclassified RCCs show more aggressive biological behaviour, in the form of larger tumor size, increased risk of adrenal gland involvement, direct invasion to adjacent organs, with regional and non regional lymphadenopathy.[8] On the contrary in our case, CT revealed a large peripherally enhancing mass, without any evidence of vascular invasion, or invasion into the adrenal gland or adjacent structures.There was no significant interval growth. Hence, unclassified RCC may show atypical imaging features, unlike the common renal cell carcinoma subtype, and may not show aggressive clinical behaviour.

CONCLUSION

Unclassified RCCs are very rare with usually metastatic potential and aggressive behaviour. This case is even much rarer variant of uRCC with clinically indolent behaviour. The clinical behaviours and imaging findings of unclassified RCCs are diverse, therefore the clinician should always keep uRCC as one of the differential diagnosis in such scenarios.

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