Renal Mucinous Cystadenoma Presenting as A Slow Growing Cystic Renal Lesion and Hydronephrosis

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Authors' contributions

Lee ZR performed the literature review, case discussion and writing of the manuscript. Tang YMM was involved in data collection and manuscript writing. Chen K was the urologist involved in primary care and surgical management of the patient. Shi RY was the reporting pathologist and contributed to the histological description of the lesion. Ho CM was the senior uroradiologist who reported the MRI kidneys and supervised manuscript writing.

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ABSTRACT

Primary renal mucinous cystadenoma is a benign cystic renal lesion reported in fewer than 50 cases in English literature. We present a case of a complex cystic lesion of the left kidney in a 74-year-old gentleman with a significant background of left urolithiasis and hydronephrosis. This lesion showed gradual increase in size over eleven years and resulted in gross hydronephrosis and chronic renal impairment. Latest CT and MR imaging demonstrated a Bosniak 4 cystic mass with enhancing nodules suspicious for malignancy. A radical nephrectomy of the left kidney and histopathological examination demonstrated a benign mucinous cystadenoma.

CASE REPORT

BACKGROUND

Primary renal mucinous cystadenoma is the benign counterpart of mucinous cystadenocarcinoma, reported in fewer than 50 cases in English literature. We report the imaging findings of a complex cystic mass according the internationally accepted Bosniak criteria, highlighting its use and pitfalls. We also detail the imaging features of a mucinous cystadenoma across MRI and CT and nuclear medicine studies, and describe the histology, pathophysiology and natural progression or this rare benign condition, and management options.

INTRODUCTION

Our patient is a 74-year-old gentleman on follow-up with Urology for a progressively enlarging left upper pole cystic lesion and gross hydronephrosis. He was asymptomatic at time of presentation. He had a significant urological history of left staghorn calculus which was treated with percutaneous nephrolithotomy in 2013.

Investigations showed poor renal function. Serum creatinine was 100-130 µmol/L (normal range 45-84 µmol/L) and MAG3 renogram showed severely reduced renal function in the left kidney (Supplemental Image 1). Urinary cultures were negative and inflammatory markers were not raised.

Contrasted CT of the abdomen and pelvis showed a large 19 x 12 x 18 cm Bosniak 4 multiloculated cystic lesion with cyst wall and septal calcifications as well as areas of, thick enhancing walls and enhancing septal nodularity (Figures 1,2, Supplemental Image 2). Renal calculi, dilation of the pelvicalyceal system and calyceal calculi and cortical thinning were also seen. Contrasted MRI of the kidneys confirmed numerous septations and enhancing nodules measuring up to 8 mm (Figure 3). The cystic component showed mild restricted diffusion with mildly raised DWI signal and low ADC (Supplemental Image 3).

Review of the CT from 11 years ago showed dilated renal calyces indistinguishable from underlying focal cystic lesion, which was at most a thin-walled lesion with no enhancing soft tissue or nodularity Bosniak 2F (Figure 1).

Though initially asymptomatic, he eventually developed intermittent flank pain and gross haematuria for 6 months. The case was discussed in a clinicoradiological Urological round and recommended for surgery in view of risk of malignancy.

Intraoperative findings confirmed a large cystic lesion arising from the left kidney adherent to the adjacent small bowel and pancreatic tail. A radical left nephrectomy was performed (Figure 4). Peritoneal lavage was performed after inadvertent rupture of the cyst.

The patient was subsequently discharged and remained well at last follow-up.

Histological features

Histology showed two dominant cyst locules containing mucoid and haemorrhagic material. The cyst wall was lined by a monolayer of columnar cells containing apical mucin (Figure 5). Abundant extracellular mucin with debris, fibrinous exudate and sheets of mixed acute and chronic inflammatory cells were seen. There were areas of pseudostratification with nuclear enlargement and nuclear hyperchromasia but no high-grade dysplasia or malignancy was seen. What appeared to be enhancing mural nodules on CT and MR were seen to be foci of inflamed granulation tissue. Immunohistochemical (IHC) staining showed diffuse positive staining for CDX2, a specific marker for cells intestinal origin, and negative staining for PAX8, a marker for organs derived from mesonephric Wolffian and Müllerian duct. This is similar to previous IHC for previously reported MC, which were positive for CDX2, MUC2, and CK20, other intestinal tissue sensitive markers [1].

DISCUSSION

Etiology & demographics

Most masses from the renal epithelium are of transitional cell or squamous cell origin while a tiny minority are adenomas or adenocarcinomas [2, 3]. Mucinous cystadenoma (MC) and its malignant counterpart mucinous cystadenocarcinoma (MCC) of the renal pelvis were first described in 1985 and are exceedingly rare [4]. Only 32 cases in scattered case reports and case series were found, approximately half of which were malignant. There is a slight male predominance of 0.68. Amongst the male patients, the mean age is 57.8 years. In comparison, the disease presents slightly earlier in the female population, mean age 47.7 years (Annex 1).

Due to a strong association with recurrent stones or infection, chronic inflammation inducing intestinal metaplasia is a postulated cause of MC [5]. Other theories suggest sequestered transitional epithelium [6] or pluripotent renal pelvic epithelium [7]. Histological similarity to mucinous cystadenoma of the ovary suggests possible mesodermal coelomic epithelium origin [8]. One patient had a caecal augmentation of the bladder and MC may have arisen directly from the colonic epithelium [9]. Disproportionate incidence of horseshoes kidneys may be related to either increase propensity for urinary stones and infections or sequestered urothelium [6].

Clinical & imaging findings

Of the 32 cases of renal MC and MCC found, approximately half of the patients presented with lumbar or flank pain (n=15); others presented with a palpable mass (n=7), or with haematuria, pyuria or mucusuria (n=7). Only 6 cases (18.8%) were asymptomatic.

Two main radiographic features are described in the literature: (i) calyceal calculi with severe hydronephrosis/dilated calyces and cortical thinning, or (ii) focal cystic mass lesion, usually Bosniak 2F – 4 (Annex 1). The focal cystic lesions may be unilocular or multilocular, demonstrate mural calcifications and mural/septal thickening, and occasionally demonstrating enhancing solid nodules. Of the few cases with MRI, signal characteristics lesions follow that or generic cystic lesions, e.g. low T1w, high T2w signal. No particular mention of functional signal characteristics such as DWI/ADC were described [6, 10, 11, 12]. Of note, the lesion is usually large, mostly between 6 to 35 cm. In cases of large mass or chronic severe hydronephrosis, there is significant overlap between both radiological patterns, as was the case in our patient.

Treatment & prognosis

MC is considered benign with an indolent course and may be treated conservatively with active surveillance. However, the malignant counterpart mucinous cystadenocarcinoma (MCC) is difficult to distinguish radiologically unless overt invasion or metastases is present and histological confirmation may be required to exclude malignancy. The general consensus criteria for malignancy are (i) macroscopic or microscopic evidence of invasion, metastasis or recurrence; and (ii) histological evidence of nuclear atypia. However, presence of nuclear pseudostratification, hyperchromasia and irregularity should be regarded as suspicious [13]. A few cases of synchronous malignancies, including primary renal carcinoid [14] or concurrent urinary bladder transitional cell carcinoma [15] have also been observed.

All cases in the literature were treated with surgical resection, usually radical or partial nephrectomy though two cases of cyst excision. No mention was made of ureterectomy, although it has been suggested that patients with mucusuria may benefit from additional ureterectomy to prevent theoretical risk of tumour seeding [16]. Some cases discovered to have an invasive component were also treated with adjuvant chemotherapy and/or radiotherapy [17, 18, 19]. Long term prognosis is not well described, due to the short follow-up periods of the available cases, most less than 5 years. The longest case of follow-up was 8 years [8]. Of the 16 cases with MCC, most were disease free at the last follow-up, with only two cases of recurrence [9,20].

One of these cases demonstrated widespread pseudomyxoma 1 year after surgery, and had also presented with mucusuria [20]. No follow-up was specified for the other patient with mucusuria [18].

DIFFERENTIAL DIAGNOSES

Renal cell carcinoma (RCC)

RCC is the most common malignant renal tumour, arising from renal tubular epithelium. It is usually centred in renal parenchyma rather than the pelvicalyceal system. Imaging findings are highly variable depending on the histological subtype, but may present with an exophytic renal lesion with central necrosis or cystic change. Cystic RCC may present as a cystic lesion with areas of mural nodularity or solid component, but is generally smaller than MC or MCC [21]. Renal vein thrombosis or metastatic adenopathy may be seen. MRI may be useful in evaluating enhancement, restricted diffusion and presence of microscopic fat, which may suggest RCC over MC or MCC. [22].

Renal epithelial cyst

Renal epithelial cysts are present in up to 50% incidence in patients above the age of 50 [23]. Cysts are variable in size, from a few millimetres up to 30 cm [24]. Smaller cysts are typically incidental findings while giant cysts may cause pain or present as a palpable lump. On US, uncomplicated cysts are typically anechoic with no internal vascularity. On CT, uncomplicated cysts are low density (0-20HU). On MRI, they are T2 hyperintense and T1 hypointense. Cyst complications such as haemorrhage or infection may mimic cystic neoplasms. The Bosniak classification is a useful tool to stratify risk of malignancy based on imaging features. Thickened enhancing septa, mural nodularity and solid component are suspicious features [25]. Bosniak 1-2 lesions are typically considered low risk while Bosniak 4 lesions have an expected malignancy rate of 91-100%.

Dilated pelvicalyceal system/hydronephrosis

The most common cause of urinary obstruction are urinary stones [26], which are also frequently seen in MC. A dilated pelvicalyceal system and most stones can be seen on US, CT and MRI. On post contrast CT and MRI, delayed renal enhancement and contrast excretion may be seen. Cortical thinning may be present depending on chronicity of insult.

Xanthogranulomatous pyelonephritis

Xanthogranulomatous pyelonephritis (XGP) is an uncommon chronic inflammatory process arising from a combination of obstruction, recurrent infection and an inadequate immune response. The most common causative organisms are Escherichia coli and Proteus mirabilis [27]. XGP is more common in females and typically presents in the middle age [27, 28]. Clinically, it can present with pain, palpable mass, haematuria or lower urinary tract symptoms. Findings may be focal or diffuse. XGP may also cause perinephric collections and fistulation into adjacent structures [27]. Large staghorn renal calculi may be seen on radiographs. On US, XGP usually presents as an enlarged kidney with staghorn calculi (echogenic pelvic focus with posterior acoustic shadowing). On CT, it displays an enlarged kidney with significant stranding, dilated renal calyces and paradoxical contracted renal pelvis, causing the "bear claw" appearance. On MRI, solid component of XGP is typically T1 iso-to-hyperintense and T2 isointense to renal parenchyma, while abscesses may demonstrate T2 hyper to isointense signal, restricted diffusion and fluid levels.

Pyonephrosis

Pyonephrosis (PYO) is a special subset of hydronephrosis, a dilated/obstructed urinary system with superimposed infection and suppurative destruction of the renal parenchyma [29]. It usually presents clinically with rapidly progressive sepsis and is considered a urological emergency warranting urgent decompression and antimicrobial therapy. Complications such as spontaneous rupture of the collecting system, abscess, fistulation and/or peritonitis may ensue if untreated. PYO tends to present in younger patients in the 4th decade of life with no gender predilection [29]. On CT, PYO demonstrates a dilated pelvicalyceal system with hyperdense fluid, urothelial thickening and significant inflammatory changes. Obstructing calculi or masses are frequently seen. On US, the dilated renal system may demonstrate dilated pelvicalyceal system with internal debris [30]. MRI may demonstrate a lower ADC value compared to hydronephrosis [31].

TEACHING POINT

Mucinous cystadenoma of the renal pelvis is the benign counterpart of a mucinous cystadenocarcinoma, and should be suspected in patients with massively dilated pelvicalyceal systems or gradually enlarging cysts with suspicious features such as mural nodularity or solid component. Serum tumour markers (CEA, CA 19-9) may be useful in raising suspicion of malignant transformation even if radiographic features are otherwise unassuming.

QUESTIONS

1. Which were the features of this lesion qualifying it as a Bosniak 4 lesion?
   a. Thickened enhancing septa on CT and MRI (applies)
   b. Mural nodules on CT and MRI (applies)
   c. Hypodense content on CT and T2 hyperintense content on MRI
   d. Posterior acoustic shadowing on US
   e. Restricted diffusion on MRI

Bosniak system classifies renal cystic masses according to their malignant potential, describing features on contrasted CT or MRI. Our patient's cystic lesion showed enhancing thickened septa (>3 mm) with mural calcification and enhancing soft tissue component and mural nodularity. Hypodense content on CT and T2 hyperintense content on MRI are nonspecific features which

may be seen in hydronephrosis or simple cysts. While posterior acoustic shadowing may represent septal calcification, it is nonspecific and may also represent calculi. Restricted diffusion is not a feature of Bosniak classification.

[Contrasted CT of the abdomen and pelvis showed a large 19 x 12 x 18 cm Bosniak 4 multiloculated cystic lesion with cyst wall and septal calcifications as well as areas of, thick enhancing walls and enhancing septal nodularity.]

2. What NOT a management option for Bosniak 4 cystic lesions?
   a. Discharge (applies)
   b. Active surveillance
   c. Ablation
   d. Nephrectomy
   e. Chemotherapy

Bosniak 4 cysts have a predicted malignancy rate of 91-100% but may progress slowly. Depending on the patient's fitness of surgery, nephrectomy or partial nephrectomy may be advised in the first instance, especially if the patient is symptomatic. However, if patients are unable to tolerate or not keen for surgery, active surveillance or ablation (for smaller tumours) may be considered. If the disease is widely metastatic, the chemotherapy may required. [Though initially asymptomatic, he eventually developed intermittent flank pain and gross haematuria for 6 months. The case was discussed in a clinicoradiological Urological round and recommended for surgery in view of risk of malignancy.]

3. What are the histological features favouring mucinous cystadenocarcinoma over the benign mucinous cystadenoma?
   a. evidence of invasion (applies)
   b. marked nuclear atypia (applies)
   c. pseudostratification
   d. metaplasia
   e. mucin production

Breach of the basement membrane (invasion) is the defining feature of malignancy on histology. Nuclear pseudostratification can be found in the respiratory tract, inner ear, vas deferens, prostate gland, epididymis, and endometrium but is considered abnormal in intestinal mucosa which normally demonstrates a neat monolayer. Nuclear pseudostratification is not a defining feature of malignancy but raises suspicion for potential malignancy. Metaplasia is a transformation of one cell type to another caused by external stimulus, potentially reversible with low malignant potential and is regarded as a benign entity; Dysplasia is the abnormal growth of cells within an organ that can harbour malignant potential and is hence regarded as pre-neoplastic. Mucin may be produced by various cell types in the body and is not in and of itself a feature of malignancy. [The general consensus criteria for malignancy are (i) macroscopic or microscopic evidence of invasion, metastasis or recurrence; and (ii) histological evidence of nuclear atypia. However, presence of nuclear pseudostratification, hyperchromasia and irregularity should be regarded as suspicious for carcinoma]

4. Which of the immunohistochemical markers stain positive for renal mucinous cystadenoma?
   a. PAX2
   b. CDX2 (applies)
   c. PAX8
   d. CD10
   e. CK20 (applies)

CDX2 is specific for cells of intestinal origin, positive in intestinal metaplasia of the urothelium that gives rise to the renal cystadenoma. CK7 stains positively for epithelial cells, nonspecific and may stain positively in the gastrointestinal tract, gallbladder, genitourinary tract, breast and lung. PAX2 is IHC marker for Mullerian origin, used in endometrial carcinoma. PAX8 is an IHC marker for cells of Mullerian and Wolffian origin, positive in central nervous system, eye, kidney, thyroid gland.

[Immunohistochemical staining showed diffuse positive staining for CDX2, a specific marker for cells intestinal origin, and negative staining for PAX8, a marker for organs derived from mesonephric Wolffian and Müllerian duct. This is similar to previous IHC for previously reported MC, which were positive for CDX2, MUC2, and CK20, other intestinal tissue sensitive markers [Chable]]

5. How do patients with renal mucinous cystadenoma present?
   a. Abdominal mass
   b. Flank pain
   c. Mucosuria
   d. Asymptomatic
   e. All of the above (applies)

While most patients in the literature are either present with pain or a palpable mass, all of the findings have been described as presentations for this entity. Patients may also represent with infective symptoms or acute renal failure. [Of the 32 cases of renal MC and MCC found, approximately half of the patients presented with lumbar or flank pain (n=15), while the next most common presentations were that of a palpable mass (n=7) or with haematuria, pyuria or mucusuria (n=7). Only 6 cases (18.8%) were asymptomatic.]

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FIGURES

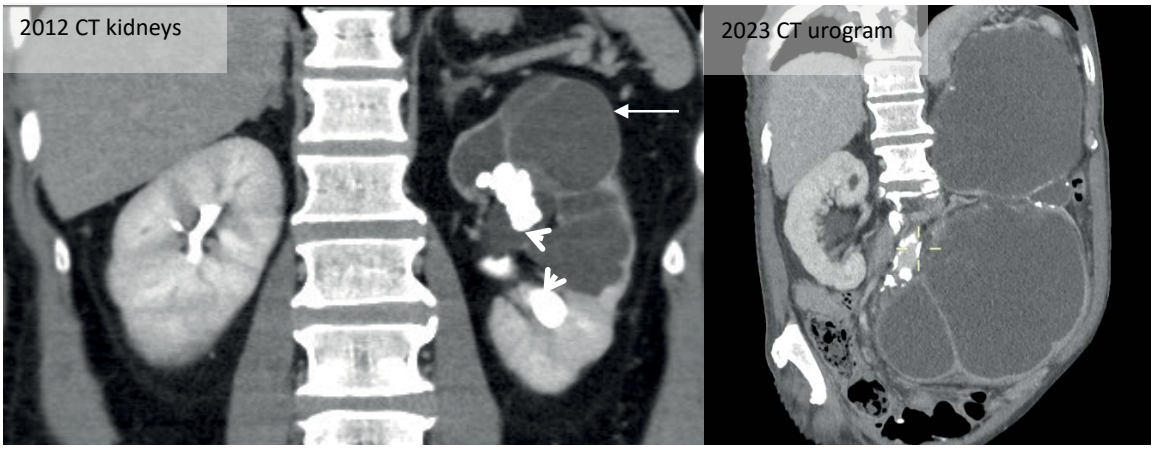


Figure 1: Post contrast CT kidneys from 2012 (a) and contrasted CT urogram from 2023 (b), both in coronal plane.
(a) Multiloculated cystic structure at the left upper renal pole (white arrow), indisguishable from dilated renal calyces in the upper and mid poles. A few renal calculi are seen (white arrowheads).
(b) New or larger left renal multiloculated cystic lesion, which demonstrates mural calcifications.

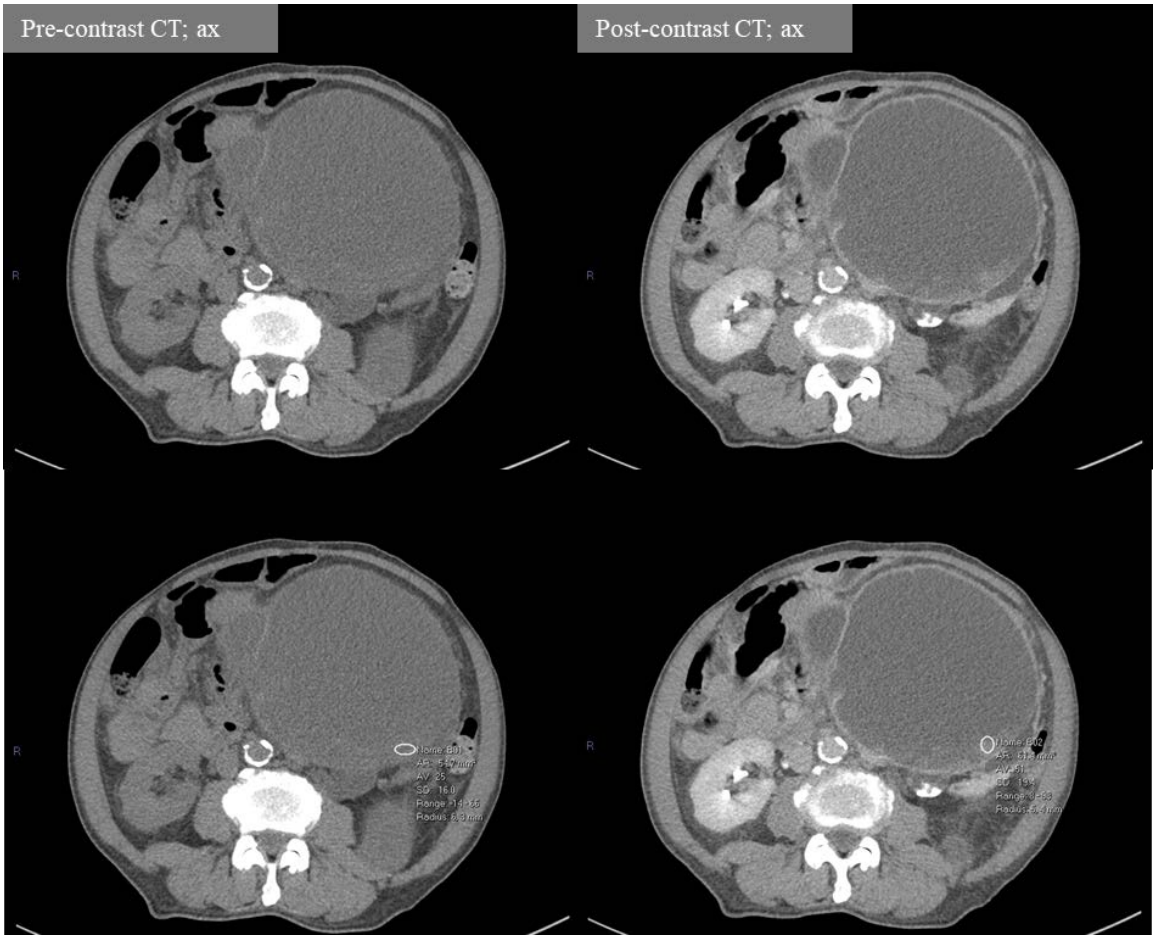


Figure 2: CT urogram in pre-contrast (a) and post-contrast nephrogenic phase (b), both in axial plane.
Subtle ill-defined enhancement in the dependent portion of the cystic lesion, equivocal for contrast excretion vs enhancing solid nodules. Region of interest shows subtle enhancement from 25HU to 51HU (bottom left, bottom right).

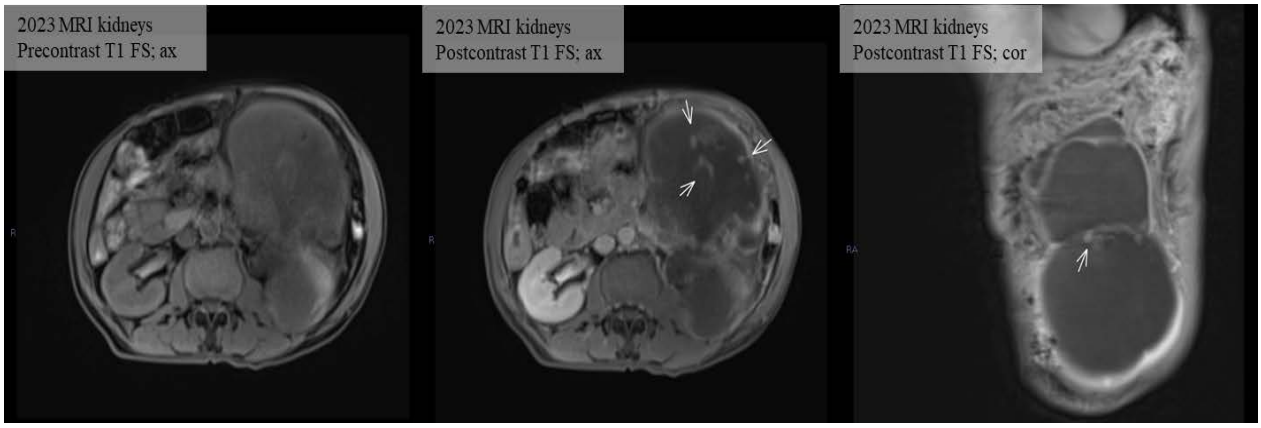
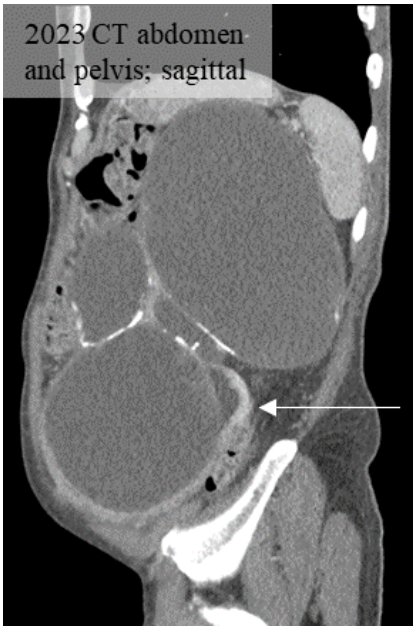
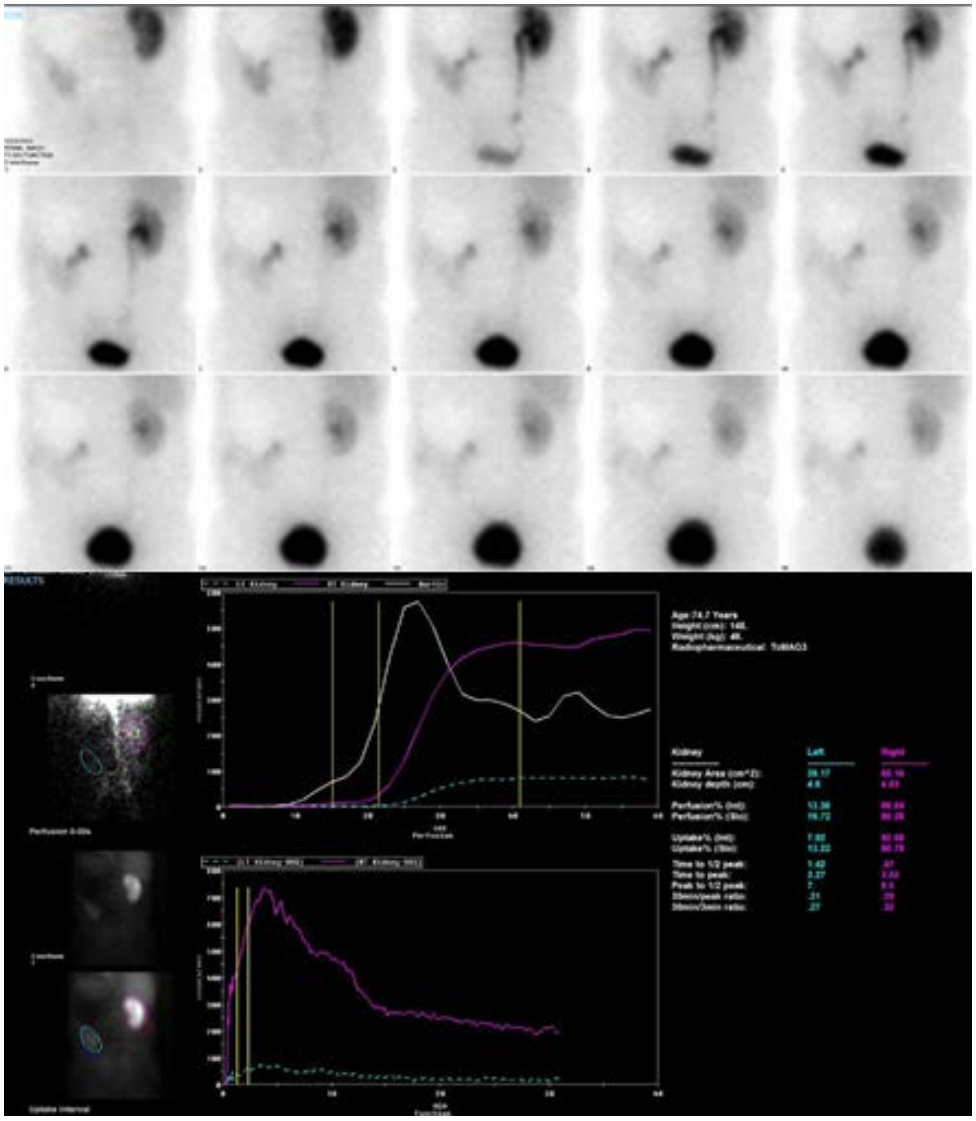


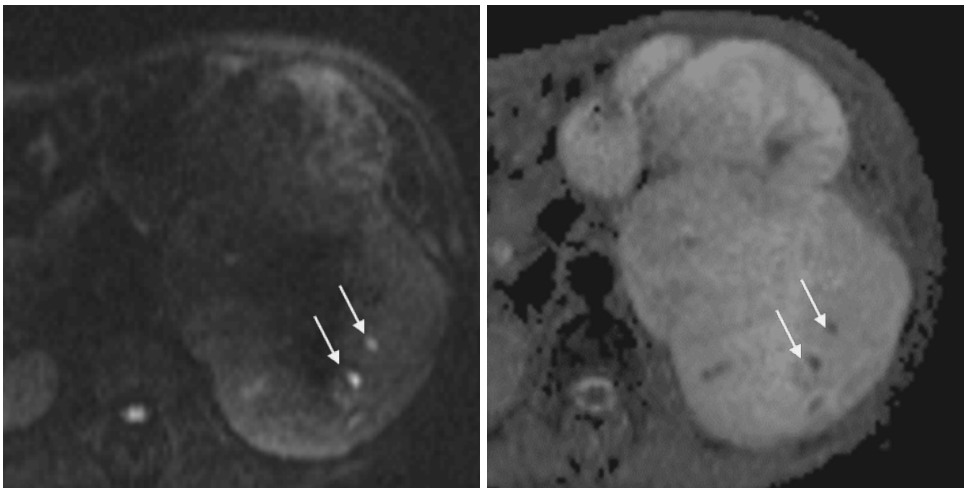
Figure 3: MRI urogram in precontrast (left) and post-contrast nephrogenic phase (middle), in axial plane. Enhancement is confirmed along the thickened septa and within the cystic lesion, some of these are curvilinear and some are nodular; already present in the pre-contrast images.



Supplemental Image 1: Post contrast CT abdomen and pelvis, in the sagittal plane. Large multiloculated cystic lesion with a thin rim of normally enhancing renal parenchyma (white arrow) demonstrating claw sign.



Supplemental Image 2: MAG3 renogram. The left kidney demonstrates negligible tracer uptake, with a severely reduced computed function of 8%.



Supplemental Image 3: MRI images, DWI, b1000 (right) and ADC (left). Mucinous component shows mild restricted diffusion, while tiny nodules within it show focal restricted diffusion (white arrows).

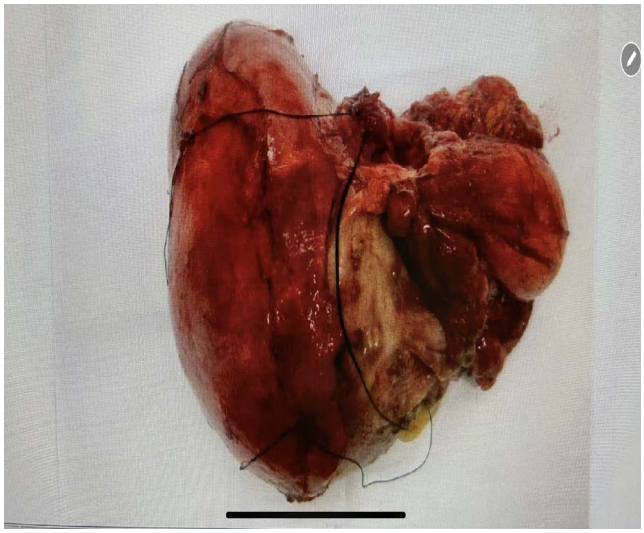


Figure 4: Intraoperative image from the left radical nephrectomy. Multiloculated cystic mass replacing almost the entire kidney.

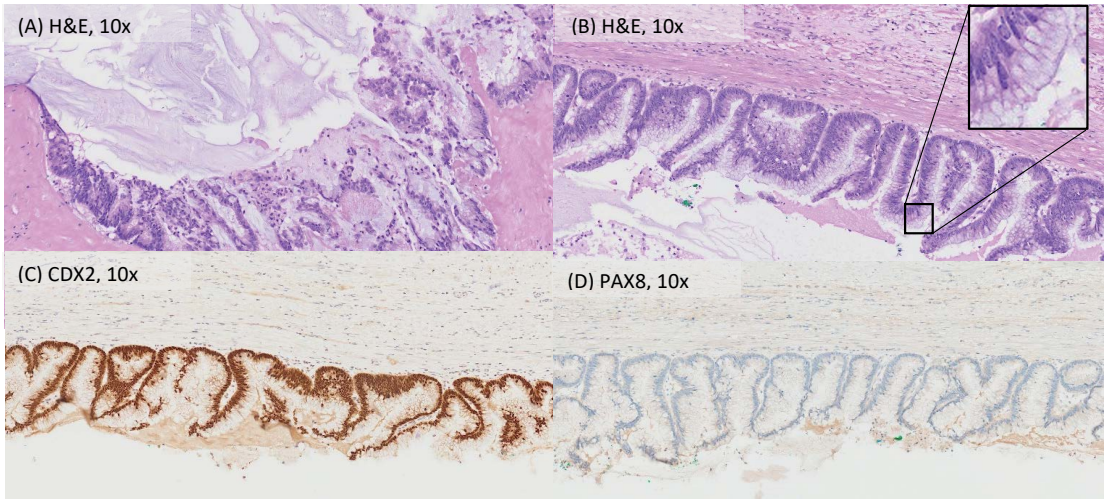
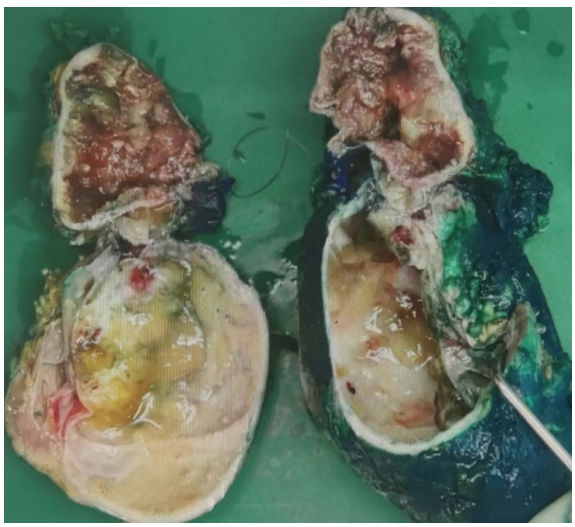


Figure 5: Histologic and immunohistochemical findings. (A) H&E, 10x magnification. Abundant fibrinous exudate and extracellular mucin with areas of patchy metaplasia. (B) H&E, 10x magnification. Replacement of normal urothelium with a monolayer of tall columnar cells containing apical mucin (see magnified inset). (C) CDX2 immunohistochemical staining, 10x magnification. Positive staining for CDX2, a specific marker for intestinal origin. (D) PAX8 immunohistochemical staining, 10x. Negative staining for PAX8, a marker for organs derived from mesonephric Wolffian and Müllerian duct.



Supplemental Image 4: Gross histological specimen with areas of mucin and nodular debris within.

SUMMARY TABLE

Aetiology	Chronic inflammation inducing intestinal metaplasia
Incidence	<1% renal tumours
Gender ratio	Male predominance, M:F=2.2
Age predilection	Male patients mean age is 57.8 years. Female patients mean age 47.7 years.
Risk factors	Urolithiasis, recurrent urinary tract infections, horseshoe kidney
Treatment	Nephrectomy
Prognosis	Long term prognosis is not well described, due to the short follow-up periods of the available cases, most less than 5 years.
Findings on imaging	Two main radiographic features described in the literature: (i) calyceal calculi with severe hydronephrosis/dilated calyces and cortical thinning, or (2) focal cystic mass lesion, usually Bosniak 2F or Bosniak 3.

Differential table

	US	CT	MRI
MC/MCC	Focal cystic lesion, with or without septation and nodularity	Two main patterns: (i) calyceal calculi with severe hydronephrosis/dilated calyces and cortical thinning, or (2) focal cystic mass lesion, usually Bosniak 2F or Bosniak 3. Obstructive calculi may be present.	Predominantly cystic, T2 hyperintense and T1 hypointense. Cystic component does not enhance but mural or solid component may demonstrate enhancement.
Renal cell carcinoma	Focal solid lesion or complex cystic lesion	Solid enhancing lesion or complex cystic lesion, Bosniak 3 or 4. Renal vein thrombosis or suspicious adenopathy may be seen.	Solid enhancing lesion or complex cystic lesion, Bosniak 3 or 4. Findings may vary but tend to be T2 intermediate or hypointense with variable T1 signal. Solid component usually enhances. High DWI and low ADC.
Renal epithelial cyst	Thin-walled anechoic cysts from a few millimetres to giant cysts up to 30 cm. Complicated cysts may demonstrate internal echoes or mural thickening.	Uncomplicated cysts are typically low attenuation, unilocular and thin walled. Complicated cysts may demonstrate wall thickening or hyperdense content.	Uncomplicated cysts are T2 hyperintense, T1 hypointense. Complicated cysts may demonstrate T1 hyperintensity or fluid levels.
Hydronephrosis	Dilated pelvicalyceal system, anechoic. May demonstrate cortical thinning depending on chronicity.	Dilated pelvicalyceal system of cystic/ plasma attenuation (HU 0-20). May demonstrate cortical thinning depending on chronicity. Calcified stones may be better assessed on CT. Delayed renal enhancement and contrast excretion.	Dilated renal pelvis with high T2 signal. Stones may manifest as signal voids in the renal pelvis or ureter.
XGP	Enlarged kidney with gross distortion of the normal renal architecture. Staghorn calculi may be seen as large amorphous echogenicity with posterior acoustic shadowing in the renal pelvis.	Multiple rounded regions of low attenuation representing dilated renal calyces are seen radiating from a contracted renal pelvis. Staghorn calculi. Perirenal inflammation presents as a thick rim of soft tissue.	Perirenal inflammation is seen as hyperenhancing thick rim around the kidney. The solid component of XGP is typically T1 iso-to-hyperintense on T2 isointense to renal parenchyma. Abscesses may demonstrate T2 hyper to isointense signal and restricted diffusion. Fluid levels may be seen.
Pyonephrosis	Dilated pelvicalyceal system, with internal echoes.	Dilated pelvicalyceal system, with higher attenuating fluid. Inflammatory changes of fat stranding. Complications such as fistulisation, perinephric abscess or peritonitis may occur.	Dilated renal pelvis with low ADC signal within

ANNEX 1

Author, year	Age	Gender	Renal abnormality / urolithiasis / HN	Symptoms	Tumour markers	Radiological features	Classification as per Bosniak 2019	Treatment	Histology	Follow-up
Zeng, 2023	55	Male	NS	Asymptomatic	Elevated CEA, CA 19-9 and CA 72-4	Bosniak 2F cyst measuring 10.0 × 8.2 cm in 2019 with partial cyst wall calcification increasing in size over serial CTs	2F	PN followed by RN and adjuvant chemotherapy and radiotherapy	MCC	No recurrence at 30 months
Nate, 2023	72	Female	NS	NS	NS	CT: gradually increasing cystic lesion with calcified septa. Bosniak 2F MRI: predominantly cystic nodule with enhancing mural nodule, Bosniak 2I	2I	Nephrectomy	MC	No recurrence at 18 months
Liu, 2022	52	Female	NS	Intermittent abdominal pain	NS	CT: multiple left renal cysts, the largest measuring 12 cm with thickened cyst walls and mural calcifications, Bosniak 2I	2I	Nephrectomy	MC	No recurrence at 8 years
Kalantari, 2021	66	Male	NS	Left flank pain	NS	US: 6 cm complex cystic mass with irregular septa in the lower pole of the left kidney (previously simple benign cyst with delicate septa). CT scan showed the same results plus calcification. Bosniak 2F. No extracystic extension, LN or mets	2F	RN	MC	No recurrence at 8 months
Tamsin, 2020	65	Female	Bilateral nephrosclerosis	Acute renal failure, night sweats, smelly urine	Normal 8 years prior	CT: Bosniak 2F cyst measuring 16cm multiple minimally thickened septa with thick calcifications. MR: Native T1w hyperintense, small enhancing mural nodule. HN from mass effect.	2F	RN	MCC	Lost to follow up
Li, 2020	66	Male	Urolithiasis, HN	Fever, flank pain	Elevated CEA and CA 19-9	CT: multiple renal pelvic calculi, stenosis of the ureter, severe HN and cortical thinning	NA - presumed HN	PCN (originally diagnosed as pyonephrosis, drained gelatinous material), followed by RN and uretectomy.	MCC	No recurrence at 12 months
Xiang, 2017	75	Male	Urolithiasis	Intermittent dull pain	NS	US: renal calculus with severe HN. CT: bilateral calyces, HN, thinned overlying cortex, multiple renal calculi	NA - presumed HN	Nephrectomy	MC	No recurrence at 13 months
Joseph, 2016	51	Male	Horseshoe kidney	Abdominal pain, palpable abdominal mass, haematuria along with passage of mucoid material in the urine	NS	CT: hydronephrotic right moiety, with grossly thinned out renal parenchyma and fusion of both lower poles, 15cm hypodense mass	NA - presumed HN	Nephrectomy and chemotherapy	MCC	NS

Lai, 2016	40	Male	No calculi	Intermittent dull lumbar pain.	CEA elevated, CA19-9 upper limit of normal	US: solid cystic mass 9.0 x 6.8 cm. CT: cystic mass with thin walls with no definite enhancement. PET CT: no significant FDG uptake	I with haemorrhage	RN followed by chemotherapy	MCC	No recurrence at 14 months
Kim, 2015	55	Male	NS	Asymptomatic	Not done	US, CT and MRI: 5.5 cm Bosniak 2I hemorrhagic cyst with a small amount of irregular enhancing septa. No LN or mets.	2I	PN	MCC	No recurrence at 12 month
Han, 2015	50	Male	Chronic urolithiasis	Flank pain after fall	NS	CT: large calculus with severe HN and cystic lesion with irregular septa	2I	Nephrectomy	MCC	No recurrence at 20 months
Mitome, 2015	45	Male	Horseshoe kidney	Palpable mass		CT: gradually enlarging Bosniak 2F unilocular cystic lesion from 8 cm to 16.5 cm in 3 years with development of HN. MR: low T1w high T2w signal, no evidence of malignancy	2F	Cyst excision after aspiration of mucinous material and development of left HN	MC	No recurrence at 6 months
Patel, 2014	45	Female	Urolithiasis	Dull abdominal pain	NS	Multiphasic CT: 17.4 x 14.0 x 7.6cm multicystic mass with calcification, no significant nodular or septal enhancement. Thinned overlying parenchyma.	2F	Nephrectomy	MCC	No recurrence at 6 months
Chable, 2013	45	Male	Recurrent left pyelonephritis	Recurrent left acute pyelonephritis	NS	CT: left HN without neophrolithiasis	NA - presumed HN	Nephrectomy	MCC	No recurrence at 64 months
Sonmez, 2013	67	Male	No calculi	Intermittent left flank pain	NS	No imaging specified	NA	Nephrectomy	MCC	No recurrence at 6 months
Chable, 2013	64	Male	Urolithiasis	Palpable mass	NS	CT: right hydronephrosis and nephrolithiasis	NA - presumed HN	Post-mortem	MC	Post-mortem (death related to myocardial infarction)
Chable, 2013	54	Male	Urolithiasis	Palpable mass	Slightly elevated CEA	CT: 20 cm multiloculated cystic mass with thin septae with a minor solid component and calcifications.	2I/4	Nephrectomy	MC	No recurrence at 24 months
Tepeler, 2011	60	Male	NS	Asymptomatic, incidental microscopic haematuria	NS	CT: Bosniak 2F 7 cm multilocular cystic lesion from renal pelvis, containing with enhancing septa and calcified walls	2F	RN	MCC	No recurrence at 24 months
Raphael, 2011	56	Male	Urolithiasis	Right flank pain	Normal CEA postoperatively	CT: HN of the right kidney, staghorn calculi and dilated ureter	NA - presumed HN	Nephrectomy and adjuvant radiotherapy	MCC	Recurrence at 1 year in the spine and lymph nodes
Fareghi, 2009	45	Male	Urolithiasis	Left flank pain, abdominal pain	NS	CT: 30 x 25 x 20 cm lobulated multicystic thin wall mass in the left kidney with thick septation and solid component without marked enhancement and some large stone. IVP: non-functional left kidney.	4	Nephrectomy	MCC	NS
Rao, 2009	52	Male	Urolithiasis	hematuria, mucusuria, abdominal flank pain, nausea/ vomiting	NS	CT: 35 cm multiloculated thin walled cystic renal mass involving the left kidney, with some thick septa and solid component	2I/4	Nephrectomy. No adjuvant treatment.	Borderline MCC. MC with intraepithelial foci of carcinoma	Recurrence at 1 year presenting with pseudomyxoma peritone2

Kumar, 2009	63	Male	Urolithiaisis	Dysuria, right abdominal lump	NS	CT: cystic enlargement of the right kidney with presence of multiple calculi in the dilated renal pelvis. Non-functional right kidney	NA - presumed HN	RN	MC	NS
Kawahara, 2009	50	Male	NS	Asymptomatic	NS	CT: predominantly cystic lesion with 2 cm solid component	4	RN	Carcinoid tumour with MC	NS
Gangane, 2008	65	Male	Urolithiaisis and urinary tract infection	Pain in the abdomen, weakness and loss of appetite, tender lump in the left flank	NS	US: enlarged left kidney thought to be pyonephrosis, left lower calyceal calculus dilated proximal left ureter	NA - presumed pyonephrosis	Nephrectomy	MCC with background pyonephrosis	No recurrence at 8 years
Gangane, 2008	35	Female	NS	Intermittent abdominal pain, lump in the left upper abdomen	NS	No imaging specified	NA	Nephrectomy	MC	No recurrence at 2 years
Charfi, 2008	31	Female	NS	Right flank pain, hematuria	NS	US and CT: multiloculated cystic mass of the right kidney measuring 13 x 7x 6 cm	Probably 2F	Nephrectomy	MC	NS
Akan, 2005	27	Female	Horseshoe kidney	Asymptomatic	NS	US and CT: unilocular cyst with no calcification or evidence of malignancy	Probably I	Cyst excision after aspiration of mucinous material	MC	No recurrence at 22 months
Takashi, 2003	53	Female	Recurrent urolithiasis	Asymptomatic	NS	US: unilocular 2 cm renal cyst with interval growth to 2.7 cm in 6 months. CT: a solid nodule protruding from the cystic lesion and mural calcification. MR: enhancement of the solid nodule	4	Nephrectomy	Carcinoid tumour with MC	No recurrence at 21 months
Park, 2002	79	Male	Urolithiasis and pyelonephritis	Fever, right flank pain, nausea	NS	CT: dilated renal calyces, calculus, surrounding inflammation	NA - presumed HN	Nephrectomy performed for presumptive xanthologranu-lomatous py-elonephritis	Renal pelvis villous and mucinous adenoma	NS
Yi, 1999	38	Female	TB cystitis and later caecal augmentation of the bladder	Painless haematuria	NS	CT: gross hydronephrosis, suspicious soft tissue in the dilated renal pelvis and multiple calcific foci	NA - presumed HN	RN and TURBT	MCC	Recurrence at the bladder within 3 months
Toyoda, 1997	69	Male	NS	Dysuria	NS	Radiograph: coarse calcifications thought to be stones. CT: ovoid cyst with nodular calcification	2F	Nephrectomy	MCC	No recurrence at 2 years
Ross, 1985	59	Female	Horseshoe kidney	Abdominal pain	NS	7 cm cyst	NA	PN	MC	No recurrence at 4 years

KEYWORDS

Trigeminal neuralgia; MRI; Meckel's Cave; Neurovascular conflict; Radiosurgery