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# ORIGINAL ARTICLE OF RENAL ANGIOMYOLIPOMAS: SYNDROMIC ASSOCIATION, RADIOLOGICAL DIVERSITY AND MANAGEMENT

Radiology

**Geetika Sindhwani** Assistant Professor, Pramukh Swami Medical college and Shree Krishna Hospital, Karamsad, Anand, Gujarat.

**Abhinav Jain** Resident, Muljibhai Patel Urological Institute, Nadiad, Gujarat.

**Manali Arora.** Senior Resident, Pramukh Swami Medical College and Shree Krishna Hospital, Karamsad, Anand, Gujarat.

**Viral Patel** Associate Professor, Pramukh Swami Medical College and Shree Krishna Hospital, Karamsad, Anand, Gujarat.

**Shabnam Bhandari** Grover. Professor, V.M.M.C. and Safdarjung hospital, New Delhi.

**Anju Ranga** Senior Resident, V.M.M.C. and Safdarjung Hospital, New Delhi.

## ABSTRACT

**Introduction:** Angiomyolipoma (AML) is a rare mesenchymal renal tumour with 80% sporadiac occurrence and 20% syndromic association with tuberous sclerosis complex (TSC). This study aims at analyzing the spectrum of Renal Angiomyo-lipoma, their vivid presentations, and associated complications which are diagnosed using Ultrasonography and Computed Tomography thereby, facilitating treatment and determining prognosis.
**Material** The above mentioned study was conducted in Department of Radiodiagnosis, VMMC and Safdarjung Hospital. Total of 10 patients with Ultrasound and CT suggestive of renal angiomyolipoma were included in study.
**Results:** Mean age of presentation was 33.5±7 (28-40) years in association with Tuberous Sclerosis. All the patients of AML with associated Tuberous Sclerosis were females. 30% of all patients (3/10) presented with hypovolemic shock. Lesions were multiple and bilateral in 40 % (n=4) of patients.
**Conclusion :** Considerable amounts of fat in most lesions, facilitate diagnosis using CT or MRI. Tuberous sclerosis is associated with female predilection, multiplicity, bilaterality, more bleeding propensity and heterogeneity of AML lesions.

## KEYWORDS

Mesenchymal Tumor Kidney, Wunderlich's Syndrome, Tuberous Sclerosis, PEComa.

### Introduction

Angiomyolipomas are benign lesions composed of variable amounts of fat, smooth muscle and thick-walled, inelastic blood vessels. They usually occur spontaneously in the general population with female predominance during their fifth decade (Jinzaki M et al 2014, Harreit J et al 2007 and Lai HY et al 2006). In patients with tuberous sclerosis complex, presenting clinically with triad of low intelligence, epilepsy and adenoma sebaceum, these frequently occur at a much younger age and manifest as multiple bilateral lesions, with an incidence of 50–80% (Jinzaki M et al 2014). They can also be rarely seen associated with other phacomatosis like neurofibromatosis and autosomal dominant polycystic kidney disease (Kumar S et al 2015)..

Around 70-80 percent of small sized lesions (< 4 cm in size) are asymptomatic and are only incidentally detected (Kumar S et al 2015). Larger lesions (> 4 cm) exhibit symptoms that include fever, nausea, pain, vomiting, palpable mass, hypertension, hematuria, anemia, and shock in around 80-85% cases (Kumar S et al 2015). Most worrisome concern for surgeon is risk of aneurysm formation and abdominal hemorrhage. Risk of hemorrhage is related to size of the tumor (significantly higher in lesions greater than 4 cm in diameter), pregnancy and association with tuberous sclerosis (Nelson CP 2002 and Steiner MS 1993). Retroperitoneal hemorrhage (Wunderlich syndrome) is seen in around 50% of patients with larger tumor (Nelson CP 2002 and Steiner MS 1993).

Pathologically, Angiomyolipoma (AML) consists of perivasculare-pithelioid cells (cells which are found surrounding blood vessels and which resemble epithelial cells), thus including them in family of PEComa. So, show immunoreactivity for both melanocytic [Human melanosome B (HMB)-45 and/or Melan-A] and smooth muscle (smooth muscle actin (SMA) and/or desmin) markers (Moriya K et al 1999 and Fujii Y et al 1995).

Appearance of lesion on ultrasound is variable depending on the proportions of contents and presence of hemorrhage. Classical angiomyolipoma appears as a ball shaped, cortical, highly reflective mass, more echogenic than renal sinus fat. Tumors with a greater proportion of muscle, and those with hemorrhagic and necrotic components, may not be echogenic and can be easily confused with renal cell carcinoma. Presence of cystic areas, hypoechoic rim and calcification favour RCC over AML. Moreover, posterior shadowing if seen favors AML. In spite of these differentiating features, a confident diagnosis of a classic angiomyolipoma requires the identification of lipid component using CT or MRI [Jinzaki M et al 2014, Fujii Y et al 1995].

CT has excellent sensitivity, specificity, positive and negative predictive values regarding AML and other renal masses. CT usually demonstrates a fatty mass intermixed with areas of increased tissue density, although the amount of fat present is variable and it can even be absent. Areas of -10 HU or lower in lesion are generally considered diagnostic of macroscopic fat (Raghavendra BN et al 1983 and Bosniak MA 1988).

MRI, being radiation free with an added advantage of high resolution and easy identification of lesion components, precludes use of nephrotoxic contrast media in patients with compromised renal function. Simple T1-weighted images with and without fat suppression and chemical shift imaging makes the diagnosis of lipid content easy to detect (Israel GM 2005).

This study aims at analyzing the spectrum of Renal Angiomyo-lipoma, their vivid presentations, extent of disease and further associated complications which are precisely diagnosed using Ultrasonography and Contrast enhanced CT thereby, facilitating treatment and determining prognosis.

### Review of Literature:

Generally seen in the cortex, AMLs are composed of adipocytes,

abnormal vasculature, and smooth muscle cells and the lesions are mostly seen in the cortex of the kidney[12].The symptomatology can include haemorrhage which is directly related to the size of the tumour and vague and dragging pain due to mass effect and obstruction causing hydronephrosis. Acute bleeding is treated with super-selective angio-embolization followed by partial nephrectomy which is preferred (Bissler JJ 2004 and Nelson CP 2002).There is some potential of metastasis (Pea M 1998 and Al-Saleem T 1998).

It is supposed to originate from perivascular epithelioid cells (Bissler JJ et al 2004). Patho-physiologically there are two types of AML: classic renal and a second type of AML containing, perivascular epithelioid cells, making it more aggressive (Bissler JJ et al 2004, Eble JN et al 1998, Oesterling JE et al 1986, Pruijm MT 2003 and Steiner MS 1993).

CT is the imaging of choice with presence of fat within mass as the diagnostic feature (Bissler JJ et al 2004. Calcification goes against its diagnosis (Lemaitre L et al 1997). Renal biopsy is contraindicated. Oesterling et al have proposed a treatment protocol based on size and symptoms of AML (Oesterling JE et al 1986). Patients with TSC and an AML >40 mm in diameter can complicate (Bissler JJ et al 2004, Eble JN et al 1998, Oesterling JE et al 1986, Pruijm MT 2003 and Steiner MS 1993).

**Material and methods:**

The above mentioned study was conducted in Department of Radiodiagnosis, VMMC and Safdarjung Hospital. Total of 10 patients with Ultrasound and CT suggestive of renal angiomyolipoma who came to our institution from 01.02.2010 to 31.01.2017 were included for the study.

**Inclusion criteria**

Patients with imaging findings suggestive of Angiomyolipoma.

**Method**

After having obtained informed consent from the patient, a detailed demographic data was obtained in the form of patient name, age, sex and contact details. After that, the detailed past and present history of the patient was sought after investigating the patient radiologically.

**Imaging protocol**

Ultrasound was done using LOGIQ P5 machine along with Colour Doppler. Contiguous helical MDCT sections of abdomen and pelvis were taken from the domes of diaphragm to ischial tuberosity. After intravenous administration of 100 ml of non-ionic iodinated contrast medium (300 mg iodine/ml Iohexol), 7.5 mm contiguous helical MDCT sections were obtained in arterial phase (at 25-35 seconds) and venous phase (at 70 seconds). Few 7.5 mm thick delayed helical MDCT sections will also be taken to look for excretion of contrast from kidney into pelvi-calyceal system and bladder.

In patients with suspected tuberous sclerosis, CT scan of head was acquired to rule out sub-ependymal tubers.

Patient with no clear fat content and poor renal function sequences of MRI were taken including T1W without and with fat saturation and chemical shift imaging.

**Radiological assessment**

On ultrasonography, renal lesion was evaluated for size (<4 cm or > 4cm), echo-texture, single or multiple, unilateral or bilateral, hydronephrosis, aneurysms, intra-renal, intra-peritoneal and retroperitoneal hematoma.

On CT, exact lesion size, location, attenuation, lesion heterogeneity, fat attenuation foci, involvement of pelvi-calyceal fat, hydronephrosis, enhancement of residual renal parenchyma, renal function on delayed scan, portion of residual renal parenchyma, aneurysm formation, any signs of rupture and associated complications were seen.

MRI, note was made of any extracellular of intracellular fat based on imaging findings on T1W and chemical shift imaging.

**Statistical Evaluation**

The obtained data was statistically evaluated using appropriate percentages using SPSS software. P value was calculated with 20 % as the power of the design and 80% being taken as the confidence interval.

**Results:**

**1) Demographic Profile**

Mean age of presentation was 33.5±7 (28-40) years in association with Tuberous Sclerosis, group A and 40.7 ±6 (32-56) years in normal population, group B. Females formed a majority of the population (70%) (n=7). The female: male ratio in sporadic AML was 1:1 while all the patients of AML with associated Tuberous Sclerosis were females.

**2) Symptomatology**

50% (n=5) patients were asymptomatic. Out of these, n=3 (60%) had lesions detected on USG for routine health checkup, while the rest were cases of Adenocarcinoma Colon and Lung with incidentally detected AML.

60% of symptomatic patients (3/5) and 30% of all patients (3/10) presented with hypovolemic shock.

**3) Characteristics of the mass**

Total of 13 renal units were involved (Bilateral Tumours in 3 cases with history of prior simple nephrectomy for large AML on one side in one of the patient). Lesions were multiple and bilateral in 40 % each (n=4) of patients. Size of the lesions ranged from 1.2 to 21 cm with a mean of 8.2 ±0.2 cm. 10 renal units (76.9%) had lesions greater than 4 cm. Approximately 46.1 % (6/13) of the units had aneurysmal dilatation of the vessels. 38.4 % (5/13) and 23.0% (3/13) patients had presence of intrarenal and retroperitoneal haematoma respectively.

**4) Association with Tuberous Sclerosis**

N=4 (40%) patients had associated features of tuberous sclerosis (Group A) including Adenoma Sebaceum (n=4) (40%), Cortical Tuber on CT (n=2) (20%) and Sub-ependymal lesion in third ventricular region (n=1) (10%). Rest of findings and their statistical significance are tabulated in Table 1.

**5) Radiological findings**

**A) Ultrasound findings**

Ultrasound findings were highly suggestive of renal angiomyo-lipoma in 9 of our patients (90 %). All of these had characteristic hyperechoic components with associated post-acoustic shadowing. Aneurysm formation was easily detected in Ultrasonography with associated Ying yang phenomena on Doppler scan. Ultrasound findings in few patients are shown in Figure 1.

In one patient, due to extensive retroperitoneal hemorrhage renal outline could not be delineated.

**B) Computerized Tomography (CT) findings**

CT was able to demonstrate fat in all patients with demonstration of fat densities (-10 HU or less) facilitating diagnosis of AML confirming 100 % sensitivity. Exact lesional delineation, extent, demonstration of aneurysm (size of aneurysm and propensity for aneurysmal rupture like teat formation) and hematoma were easily seen in all cases on CT scan. There is additional advantage in delineating vascular supply of lesion and aneurysms and amount of residual normal renal tissue providing necessary therapeutic planning. Hydronephrosis was seen in 4 large lesions causing compression of pelvicalyceal system.

Radiological findings in Tuberous sclerosis patients are shown in Figure 2, 3, 4 and 5. CT findings in non-syndromic patients are shown in Figure 6.

**Management**

Patients with Symptomatic lesions irrespective of the size and the characteristics of the patients were offered minimally invasive surgery with a preference to nephron sparing surgery as per the feasibility. Females of child bearing age group irrespective of the size of the lesion or the symptomatology were offered surgical management. Lesions less than 4 cm were offered conservative management.

The patients with emergent presentation were managed with resuscitation and referred to other centre for trans-femoral super selective renal arterial embolization followed by evacuation of the perinephric collection.

**Discussion:**

Angiomyolipoma (AML) is a rare mesenchymal renal tumour constituting only 0.3 to 3% of all aggregated renal tumors. Renal angiomyolipoma has been known to occur in two different clinical settings: in association with tuberous sclerosis and sporadically (Jinzaki M 2014).

Out of total 10 patients, females predominated group forming 70 % of cases. A large study in Japanese population showed an incidence of

0.1% in male and 0.22% in female patients (Fujii Y 2005).Of our 10 patients, 6 (60%) had sporadic angiomyolipoma and the other 4 (40%) had associated tuberous sclerosis.

Amongst those presenting with sporadic angiomyolipomas, the mean age at presentation was 40.7 ±6 years and the female to male ratio was 1:1. Of the female patients, all were peri-menopausal with a mean age of 50.1 years. Both kidneys were equally involved. All of these sporadic patients had solitary lesion on ultrasound and CT scan. Similar findings were seen in a single centered study by Sanjay Gagoi in Indian set up. Kumar S has reported female predominance in sporadic cases in review of literature, due to estrogen and progesterone sensitivity of tumour (Kumar S 2015)

Of the patients presenting with tuberous sclerosis, one had cutaneous & ocular stigmata, 2 had subcortical tubers on brain CT scan and all had bilateral renal angiomyolipoma. All four were female patients with a mean age of 33.5 years. All four patients had bilateral multiple lesions. For one patient, unilateral nephrectomy was done earlier. Stilwell et al studied 95 patients with tuberous sclerosis and found that 47% of the patients had angiomyolipomas and among these 71% had bilateral angiomyolipoma and 87% had multiple lesions. Gomez et al revised diagnostic criteria and postulated multiple angiomyolipomas as one of diagnostic criteria for tuberous sclerosis. In our study, 4 out of 10 patients had tuberous sclerosis and all patients revealed bilateral and multiple lesions.

Casper et al also concluded similar findings in his study, with female predominant occurrence, bilaterality and multiplicity of lesions.
Grawitz gave the term angiomyolipoma in 1900 to describe this tumor for the first time based on histological findings. The incidence of this rare tumour is less than 1 % of surgically excised renal tumours.

**Presentation:**
These patients require more rigorous evaluation as angiomyolipoma can also occur in the brain, eye, heart, lung and the bone. These patients gravitate towards multifocal and bilateral disease, younger age of presentation, larger size of lesions, increased rate of lesion inflation and more symptomatic presentation. Thus, these patients require more frequent surgical management as compared to sporadic occurrence patients.

According to Gagoi, Sporadic angiomyolipoma can present in three varied clinical presentations. Most common presentation being incidentally detected asymptomatic group. Second group consisted of patients with mild to moderate symptoms due to local discomfort and gastrointestinal symptoms secondary to compression on the stomach and duodenum. The third group presents acutely with abdominal pain or shock because of massive intra-lesional or retroperitoneal hemorrhage. In our study group of sporadic patients, 5 out of 7 (71.4%) patients were asymptomatic, thus comprising major group. Out of other 2, one patient presented with chronic dragging pain and other patient came in shock and un-recordable BP due to massive intra-lesional and retroperitoneal hemorrhage.

Symptomatology of patients is related to the tumor size. In our study, 80 % of renal units more than 4 cm were symptomatic. Abdominal or flank pain (50%), and palpable mass in one patient were common symptoms. Similar results were seen in study by Gagoi. Spontaneous intrarenal or peri-renal hemorrhage extending to the retroperitoneum (Wunderlich's syndrome) is a rare presentation and is seen mainly in larger lesions. Its reported incidence is about 10% (Gagoi S 2001).

**Imaging**
Radiological imaging allows confident diagnosis of renal angiomyolipoma in bulk of cases. CT scan estimation of lipid density is almost characteristic of an Angiomyolipoma ( Bosniak MA 1988). In our study, Ultrasound could detect 9 lesions. CT could identify fat in all the patients. Findings seen were consistent with study with Gagoi.

In patients with tuberous sclerosis, lesions seen were more heterogeneous and conglomerated due to presence of more smooth muscle component and less of fat. These types of lesions may be impossible to distinguish from renal cell carcinoma pre- operatively (Lemaitre L 1997).
In our study, 6 out of 7 renal units in patients with tuberous sclerosis revealed heterogeneity of lesion with less of echogenic component on ultrasound. On CT scan, these patients showed very less proportion of fat as compared to sporadic cases.

In sporadic cases, 5 out of 6 renal units showed fat predominated lesions constituting around 80- 90 % of lesional volume. One patient revealed mild heterogeneity with more of enhancing component and around 20 % fat component, however, no other signs of tuberous sclerosis were seen in patient. In some instances, the occasional occurrence of radiologically detectable fat in other renal tumours, such as Wilms tumour and renal cell carcinoma can lead to misdiagnosis (Lemaitre L 1997).

**Complications**
Risk of spontaneous hemorrhage runs parallel with tumor size. In our study, all 3 renal units < 4 cm were asymptomatic making 100 %. Out of 10 renal units of size > 4 cm, 2 patients were asymptomatic constituting 20 %. Rest of large sized lesions (size ranging from 10 cm to 21 cm) were symptomatic including chronic pain (20 %), acute pain (30 %) and shock (30 %).

Van Baal et al took 3.5 cm as cut-off above which the risk of hemorrhage is high. On the other hand, Koh et al found that size greater than 6 cm was associated with hemorrhage and formation of pseudo aneurysm.

**Management**
Appropriately selected cases of renal AML like lesions < 4 cm in size and predominantly fatty lesions can be managed by active surveillance. Criteria for intervention in cases of Renal AML are symptomatic lesions larger than 4 cm, suspected malignancy and presence in women of childbearing age. Some authors have also added criteria's like aneurysm larger than 5 mm, concomitant TSC and poor access to follow-up or emergency care as additional contemplations for treatment. Rapamycin (Sirolimus), Everolimus and Temsirolimus are commercially available immunosuppressant used in patients with angiomyo-lipoma associated with tuberous sclerosis (Sooriakumaran P 2013)

**Conclusion:**
Because most angiomyolipomas contain considerable amounts of fat tissue, it is usually diagnosed using CT or MRI by identifying imaging features of fat cells in the mass and present with varied presentations. There is considerable association of bilaterality, multiplicity, female sex predominance, larger size, heterogeneity and propensity to bleed with syndromic AML.

**Tables:**
**Table 1: Table comparing Angiomyolipoma (AML) findings in syndromic and sporadic population patients.**

| Findings | Group A (Patients with Tuberous sclerosis) | Group B (Normal Population) | P value |
|---|---|---|---|
| Bilaterality | 4 | 0 | |
| Multiplicity | 4 | 0 | |
| Heterogeneity | 4 patients (6/7 renal units) | 1 patient, (1/6 renal units) | 0.029 |
| Size | 10-21 cm(11.9 ±0.3 cm) | 1.2-11 cm( 9.4 ± 0.4) | <0.001 |
| Propensity to bleed | 2 | 1 | 0.5 |

**Figures:**
**Figure 1: Ultrasonic depiction of angiomyolipoma: A.** Ultrasound image showing a well-defined lesion in right kidney with a globular anechoic aneurysm with Ying-yang phenomena. B. Ultrasound of patient showing heterogeneous lesion with echogenic areas in right kidney lower pole. C. Ultrasound image of 51 year old asymptomatic female patient showing echogenic lesion arising from upper pole of kidney causing mild hydronephrosis.

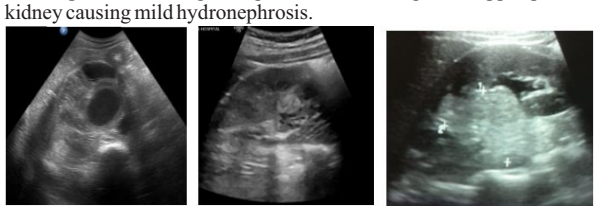


**Figure 2:** A 30 year old female presented with pain in abdomen,

chronic seizures and adenoma sebaceum. **A.** Ultrasound image showing a well-defined lesion in right kidney with a globular anechoic aneurysm with Ying-yang phenomena. **B.** Contrast enhanced CT scan axial image: There are multiple heterogeneous lesion with few fat attenuation foci in bilateral kidneys. Lesion is right kidney shows intensely enhancing aneurysm in lesion with surrounding hyperdense hematoma. The patient was managed by laparoscopic left simple nephrectomy and right partial nephrectomy. C. Head CT scan axial image showing multiple sub-ependymal calcified tubers.

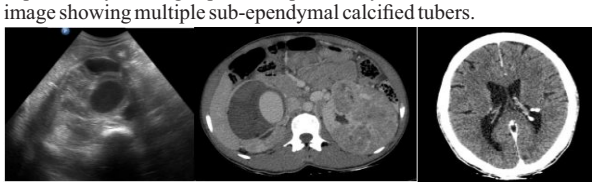


**Figure 3:** A female patient presented known case of tuberous sclerosis with chronic abdominal pain. **A** and **B**: Plain and contrast enhanced Computerized Tomography images showing right enlarged kidney with large heterogeneous lesion having few areas of fat density in right kidney and intensely enhancing globular aneurysm. There is associated surrounding non-enhancing mildly hyperdense hematoma. However, no retroperitoneal collection is seen. Rim of enhancing renal parenchyma is seen surrounding lesion. Left kidney also shows complex lesion in mid-pole. She was offered laparoscopic right simple nephrectomy in view of his symptoms.

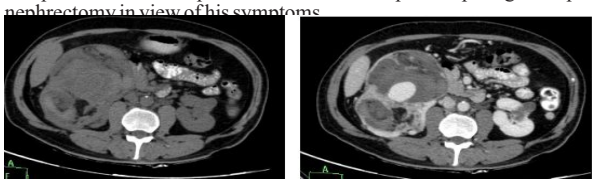


**Figure 4**: 35 year old patient presented with acute abdominal pain on right side. **A** and **B**: CT scan coronal and axial images showing bilateral enlarged kidneys with multiple diffusely distributed lesions with fat density seen within. There is small hyperdense collection in peri-renal region near superior pole of right kidney suggestive of

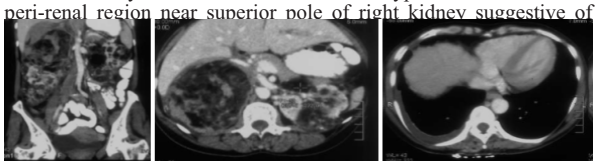


**Figure 5**: 40 year old female came to emergency department in shock. There was history of prior nephrectomy of right kidney due to bleeding in AML. **A**. Plain CT scan showing ill-defined heterogeneous lesion with hyper dense component in left kidney. There was associated peri-renal and peri-hepatic collection suggestive of acute bleeding from AML. **B** and **C**: angiography and angio-embolisation done at outside

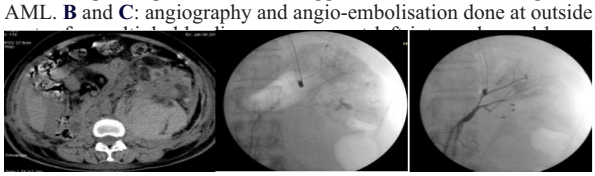
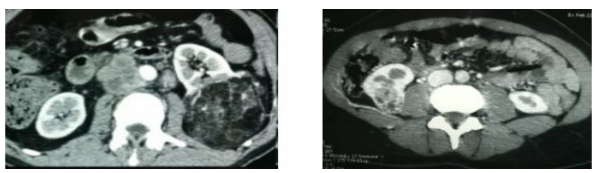
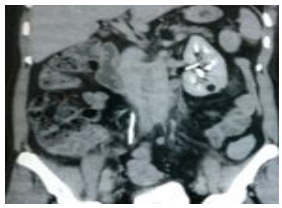
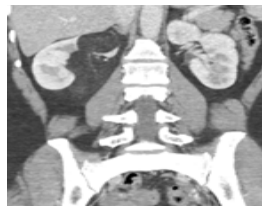


**Figure 6: AMLs in non-syndromic patients: A**.55 year old male patient with diagnosed carcinoma rectum, CT scan revealed incidental finding of heterogeneous fat attenuation lesion in mid and lower pole of left kidney with interspersed enhancing septas with few metastatic nodes in right para-aortic location. **B**. Asymptomatic female patient for health check up with ultrasound suspected of renal mass. Axial CT scan images showing heterogeneous lesion in right kidney lower-pole arising from posterior cortex with interspersed fat attenuation foci. **C**. Patient with incidental detection of small fatty density lesion in left kidney lower pole. He was managed conservatively. **D**. CT scan of 51 year old female patient showing completely fat attenuation lesion arising from right kidney upper pole and further extending inferiorly along pelvic region.







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