Multilocular cystic nephroma

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Section: Uroradiology & genital male imaging
Area of Interest: Abdomen
Procedure: Education
Imaging Technique: Ultrasound
Imaging Technique: CT
Imaging Technique: Ultrasound-Colour Doppler
Special Focus: Pathology Cysts Case Type: Clinical Cases
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Patient: 39 years, female

Clinical History:

A 39-year-old female presented with menorrhagia for 4 months. No associated dysmenorrhea or abdominal symptoms were detected. Following gynaecology review, an ultrasound of the pelvis was performed. Due to an incidental abnormal renal finding on this examination, computed tomography of the abdomen was subsequently performed.

Imaging Findings:

Ultrasound scan of pelvis revealed normal appearances of the uterus, endometrium and ovaries for a pre-menopausal patient. Sonographic examination of the kidneys is routinely performed on all pelvis ultrasound examinations at our institution. A 6.4 x 8cm complex mass was incidentally noted arising from the interpolar region of the right kidney. Computed tomography (CT) of the abdomen was recommended to further characterise this mass. A triphasic renal mass protocol CT of the abdomen demonstrated a corresponding 8cm multiseptated complex cystic mass arising from the anterior interpolar region of the right kidney, indenting the under-surface of the right lobe of the liver. There was no evidence of tumour extension or thrombus within the renal veins or IVC.

As the imaging findings could not differentiate a cystic renal cell carcinoma from a multilocular cystic nephroma, the referring urologist elected to perform a right nephrectomy. Histological analysis confirmed the diagnosis of a multilocular cystic nephroma.

Discussion:

A multilocular cystic nephroma is a rare benign cystic lesion of the kidney of unknown aetiology. Theories of its pathogenesis include dysplastic, hamartomatous, neoplastic, and dysontogenetic transformation[1, 2]. The criteria for diagnosis of multilocular cystic nephroma[3] include the composition entirely of cysts and septa, a discrete mass which is well demarcated from the non-cystic renal parenchyma and septa are the only solid portions of the tumour.

Multilocular cystic nephromas have a bimodal age and sex distribution and tend to occur in children between 3 months and 4 years of age and in adults (mostly women) between 40 and 60 years of age. The clinical manifestations of multilocular cystic nephroma are highly variable. It most frequently manifests in children as a painless abdominal mass. Adults are more likely to present with abdominal pain or hematuria, whereas a painless mass, urinary tract infection, or hypertension are less common manifestations among adults. [4]

Because neither the clinical nor the imaging features can clearly distinguish these lesions from a cystic renal cell
carcinoma, surgery—either nephrectomy or nephron-sparing surgery—is required for both diagnosis and treatment. [5]

The ultrasound appearance of multilocular cystic nephroma is typically a complex cystic mass with multiple anechoic spaces separated by hyper-echoic simple septa [6]. The renal origin of the mass can be confirmed by identification of a claw of normal renal parenchyma around the periphery.

Computed tomography (CT) findings are dependent on the size of the cyst and the amount of stromal tissue. The mass lesions will typically demonstrate well-defined margins, multicystic architecture and enhancing septations.

Findings on magnetic resonance imaging (MRI) include high T2 signal intensity within the cystic component, with low T2 signal intensity in the tumour capsule and septations. On T1-weighted sequences, variable signal intensity from the cyst contents is attributed to differing concentrations of old hemorrhage and protein [7].

Despite classical imaging findings on ultrasound and CT, the radiological appearances of a multilocular cystic nephroma are indistinguishable from a cystic renal cell carcinoma. The diagnosis must always therefore be a histological diagnosis [8]. The radiologist's awareness of and consideration of this benign entity is important particularly in patients who have a solitary kidney or are unsuitable surgical candidates.

**Differential Diagnosis List:** Multilocular cystic nephroma, Cystic renal cell carcinoma, Renal cyst

**Final Diagnosis:** Multilocular cystic nephroma

**References:**


Description: This image demonstrates a multiseptated cystic lesion arising from the right kidney.
Origin: Department of radiology, University Hospital, Galway, Ireland
Description: This image demonstrates the avascular appearance of the septations within the multiseptated renal mass. Annotation on the image refers to the proximity of the lesion to the right lobe of the liver. Origin: Department of radiology, University Hospital, Galway, Ireland
Description: Axial non-contrast enhanced CT image demonstrates a large multiloculated cystic mass arising from the right kidney. No calcification of the septations detected. Origin: Department of Radiology, University Hospital, Galway, Ireland.
Description: This axial image demonstrates enhancement of the fine septations of this cystic mass. No discrete solid nodules were identified. Origin: Department of Radiology, University Hospital Galway, Ireland.
Figure 5

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Stroma positive: Smooth muscle Actin (10 x)

Description: Histopathology of the resected nodule demonstrated multiple variable-sized cysts which were separated by septae containing fibrocollagenous stroma enriched by smooth muscle bundles. No evidence of malignancy detected. The diagnosis of MLCN was confirmed. Origin: Department of pathology, university hospital, Galway, Ireland.
Description: Histopathology of the resected demonstrated multiple variable-sized cysts which were separated by septae containing fibrocollagenous stroma enriched by smooth muscle bundles. No evidence of malignancy detected. The diagnosis of MLCN was confirmed. Origin: Department of pathology, university hospital, Galway, Ireland.