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Cystic Nephroma: An Unusual Renal Entity

KEYWORDS: Cystic nephroma, Multilocular renal cyst, MESTK

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ABSTRACT *The kidney is one of the most common sites for cyst formation in the body. The spectrum of renal cysts is wide and complex, from genetic to neoplastic in etiology, bimodal distribution in age & sex. Cystic nephroma is a rare benign neoplasm of the kidney with excellent prognosis after nephrectomy. Preoperative diagnosis of cystic nephroma is clinically and radiological difficult from other cystic renal malignancies. It is a rare benign lesion of the kidney with very few reported cases in the world literature. Even with improved precision in radiological imaging, the final diagnosis is only possible by histopathological examination of the resected specimen. This is a case report of a 30 year old female patient who presented with an incidental finding of complex multilocular renal mass. She underwent nephrectomy, and the histological findings were consistent with cystic nephroma.*

Case report

A 30 year old female patient referred with history of appendicectomy about one & half month ago to our hospital. Incidental finding during ultra sonography was a complex right renal mass. she was admitted for further management of the mass. There was no history of right renal angle pain or hematuria. Routine hematological and biochemical investigations were within normal limits. C.T. scan showed a complex multiseptate cystic renal mass of 8 cm in diameter causing severe indentation over the adjacent superior & middle calyx (Fig No.1). After preoperative work up she underwent right nephro-ureterectomy through a right sub costal incision. Renal vessels were clamped and transfixed with 2/0 silk. Right ureter was traced till crossing of gonadal vessels & divided. The specimen of kidney and ureter was sent for histopathological examination.

Drain was kept and incision closed in layers. Post-operative recovery was uneventful. She is under follow up for the last three months.

The specimen measured 12.9 x 5.6 x 6.5 cm & weighed 348 gms. On gross Examination (Fig No. 2) the upper pole was bossealated and cut section showed well circumscribed encapsulated tumour of a diameter of 7.5 cms with numerous cysts of varing size from 0.5 to 2 cms containing clear fluid. Microscopic examination showed characteristics of cystic nephroma with multiple cysts lined by flattened to cuboidal hobnail cells separated by stroma. The stroma was edematous ovarian type with lymphoplasmacystic infilterate.

Discussion

A cystic nephroma is a benign cystic lesion of the kidney of unknown etiology. Theories of its pathogenesis include dysplastic, hamartomatous, neoplastic, and dysontogenetic transformation.[1, 2]

Edmunds[3] reported the first case of cystic nephroma (CN) as cystic adenoma of the Kidney. Less than 200 cases are reported in the international literature. Multilocular cystic nephroma can be congenital or acquired.

The congenital form is seen less than two years of age with a male: female ratio of 2:1, and is generally unilateral but bilateral lesions have been reported4, 5.

In the acquired type it is seen affecting predominantly postmenopausal females[6] with a male: female ratio of 1:9[7].

Adult cystic nephroma shares the same morphologic characteristics with congenital cystic nephroma but has a different biological behavior.

Congenital cystic nephroma are sometimes associated with nodules of Wilms tumour among the benign cysts[7], and the exact etiology is unresolved.

According to classification by Eble & Bonsib[4] Cystic nephromas are thought to represent one end of a spectrum of related cystic lesions, including cystic nephroma, multilocular cyst with partially differentiated Wilms tumor, multilocular cyst with nodules of Wilms tumor, and cystic Wilms tumor[8].

WHO classification[9] -Cystic nephroma is classified as a special entity that is identical to mixed epithelial and stromal tumors (MESTK) of kidney.

The pathogenesis is based upon the influence of hormones hence common in females, in rare cases of male patients there may be history of hormone manipulation for the treatment of prostate cancer[11]. And to support the hypothesis there is presence of both estrogen and progesterone receptors[10].

Boggs and Kimmelstiel have defined certain criteria to differentiate cystic nephroma from others like polycystic disease multi cystic kidneys, simple renal cysts and cystic renal cell carcinoma[9]. The criteria are multilocular lesion, cysts lined with epithelium, cyst that do not communicate with the pelvis and normal residual renal tissue. In spite of this the final diagnosis is almost exclusively based on immunohistochemistry.

Clinically cystic nephroma usually presents as asymptomatic & incidental finding as in our case. It may present as painless flank mass or pain & hematuria[2, 8]. .

The gross specimen shows a conglomeration of non communicating cysts of varying sizes, from several millimeters to 4 cm in diameter. The cysts

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are well circumscribed by
a thick outer pseudocapsule that compresses the adjacent normal renal parenchyma.
The treatment of choice for cystic nephroma is surgical removal of kidney is correct.
Mural nodules of nephroblastoma, renal adenoma, adenocarcinoma, and sarcoma
have been known to coexist with cystic nephromas in adults.[12, 13]
In addition, diagnostic radiographic imaging (with or without percutaneous biopsies)
does not reliably distinguish malignant from benign lesions.[12]
Cystic look-alike lesions, such as cystic nephromas, multilocular cysts with partially
differentiated Wilms tumors, multilocular cysts with nodules of Wilms tumor, cystic
Wilms tumors, cystic renal adenocarcinomas, sarcomas, and mesoblastic nephromas
cannot be differentiated accurately without a postoperative tissue analysis.[2,8]
The treatment of choice in the present case was nephrectomy because of the extent of
renal involvement. Nephron-sparing surgery, however, should receive major
consideration: cystic nephromas are benign, and the possibility of a recurrence
developing after complete surgical extirpation of the neo plasm is unlikely.

Conclusion

This is a rare benign lesion of the kidney with less than 250 cases reported in the world
literature. In cases of cystic nephroma where preoperative diagnosis between it and
cystic renal cell carcinoma is impossible, hence nephrectomy is the preferred treatment.
Despite limited experience in diagnosis and treatment the need for follow is important.
Cystic nephroma also presents with no specific urinary tract symptoms like flank pain,
hematuria or hypertension and in adults may present as a incidental finding. The IVP
shows a well defined mass in a normal functioning kidney. USG shows a multi loculated
cystic renal complex mass. Color Doppler is required to differentiate between malignant
and benign lesion. Cystic nephroma are hypovascular and MRI angiography may be an
alternative tool in preoperative evaluation.

Fig No1 CT scan

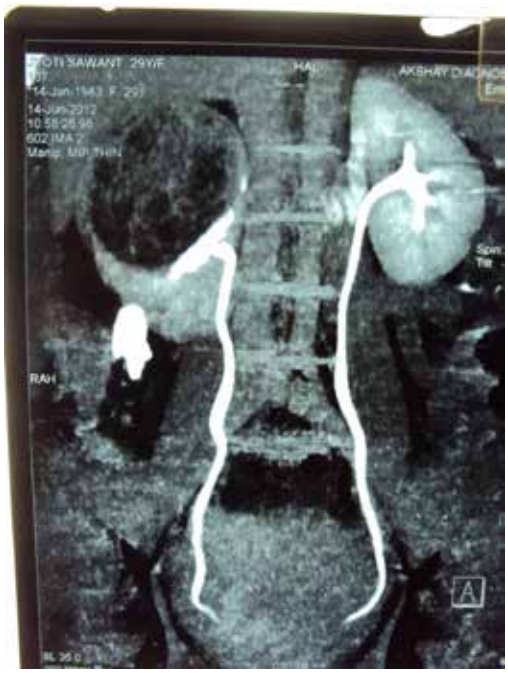

Fig No.2 Specimen

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