

Case Report-4

Cystic Nephroma: An Operative Dilemma

PARAS R KOTHARI, DINESH SARDA, GOWRI SHANKAR, RAGHUNATH DIPALI, AND BHARATI KULKARNI.

ABSTRACT

A 2-year-old male child presented with multi locular cysts in lower pole of left kidney. The diagnosis of cystic nephroma was suggested by sonography and CAT scan but established only by its pathological features.

INTRODUCTION

Cystic nephroma is an un-common, benign renal disease with unknown etiology. Origin is designated as being dysplastic or hamartomous or neoplastic? Two peaks of incidence are seen for this disease. Herniations of some cysts into urinary tract demonstrate a characteristic radiological sign.

CASE : A 2-year-old male child presented with mass in the left side of abdomen noticed by mother since last one month. There was no history of urinary or bowel complaints, fever, pain in abdomen or vomiting.

General examination revealed pallor. Blood pressure was 90/60mmHg. Systemic examination of abdomen showed a 10 x 10cms solitary mass occupying left hypochondrium, epigastrium, left lumbar and umbilical regions. It was smooth surfaced, firm to hard in consistency with restricted mobility. It was palpable bimanually and left renal angle fullness was present.

Investigations: Hb 9.7gm%. Blood urea nitrogen and serum creatinine were 7mg% and 0.5mg%, respectively. Urine routine examina-

tion and culture showed no abnormality. Ultrasonography detected a 10 x 8 cm well defined multiloculated anechoic lesion with thin septae arising from lower pole of left kidney with post acoustic enhancement. Chest x-ray showed no lesions. CAT scan confirmed the sonography findings, adding information of lesion being hypo dense containing clear fluid. No enhancement was seen on contrast.

Provisional diagnosis of multi locular renal cyst or cystic Nephroma was entertained.

Exploratory laparotomy confirmed a 10 x 8 x 5 cm huge smooth surfaced cystic renal mass arising from lower pole of left kidney. (Fig-1). There was no adhesions and lymphadenopathy. After clamping the renal vessels nephro ureterectomy was done. No additional blood supply was noticed. Postoperative period was uneventful.

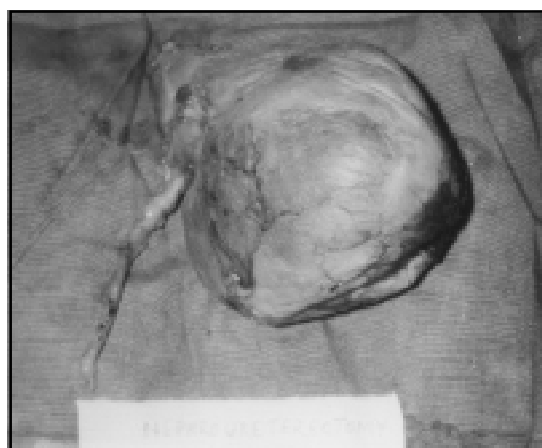


Figure-1 Photograph of gross specimen showing a 10 x 8 x 5 cm huge smooth surfaced cystic renal mass arising from lower pole of left kidney with the ureter.

Department of Paediatric Surgery, L.T.M. Medical College and General Hospital, Sion, Mumbai-400022. (INDIA)

Correspondence to: **PARAS R KOTHARI**
E-mail : drparaskothari@rediffmail.com

Gross specimen showed smooth surfaced kidney as a multiloculated cyst filled with clear fluid and no solid components. Septal thickness was 1.2mm. Cyst size varied from 2-3cms each and were non-communicating. Small remnant kidney was present at upper pole with dilated pelvicalyceal system. (Fig-2). Histopathology showed the cyst lined with columnar to flat epithelium. Ultra structure displayed cilia. Stroma between the cysts had fibroblastic tissue with dilated vessels and no metanephric blastema. Findings suggested a final diagnosis of cystic nephroma.

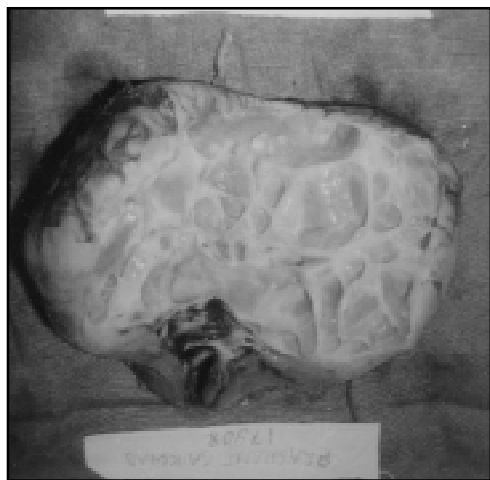


Figure- 2 Cut specimen showing multiloculated cysts containing clear fluid. Septae were 1.2 mm thick and non-communicating.

DISCUSSION

Cystic nephroma is an uncommon benign renal neoplasm. The characteristic is structurally similar to cystic partially differentiated nephroblastoma.^{1,2}

Etiology and pathogenesis of cystic Nephroma is unknown. It is debated to be neoplastic/dysplastic or hamartomous in origin. Bimodal incidence is noted. 50% occur in children < 4 years and 30% in 5th – 6th decade³. It is usually solitary and rarely multiple. Surgical excision/

histology (FNABC/Biopsy) is the only effective method to distinguish benign from malignant cystic lesion of the kidney. Some cysts can herniate into the urinary tract and can be completely excised. They are well encapsulated & non in filtrating lesions.

Cystic Nephroma having close structural similarity to nephroblastoma supports the idea of some surgeons of total nephrectomy as the treatment of choice where intra operative biopsy is ambiguous. The differential diagnosis of cystic nephroma includes cystic wilms tumour and cystic partially differentiated nephroblastoma.^{4,5,6} Nephron sparing surgery can be the therapy of choice where the lesion is well encapsulated and where the benign nature of the cyst is not doubtful. Metastasis or local recurrences have not been reported in cases of cystic Nephroma.

Few cases have been reported in the literature and the surgical treatment of nephrectomy/nephron sparing surgery is debated.

REFERENCES:

1. Okuda T, Yoshida H, Matsunaga T, Kouchi K, Ohtsuka Y, Saitou T, Horie H, Ohnuma N. Nephron sparing surgery for multilocular cyst of the kidney in a child. *J. Pediatr. Surg.* 2003;38(11):1689-92.
2. Rebassa Lull MJ, Munoz Velez D, Hidalgo Pardo F, Gutierrez Sanz – Gadea C, Mus Malleu A, Torrens Darder I, et al. Cystic Nephroma. Report of 5 cases. *Arch Esp Urol.* 2000;53(5):476-9.
3. Leva G, Annoscia S, Lozzic, Montefiore F, Di Mauro A, Geraci E, Bocca Foschi C. Cystic Nephroma: report of two cases and review of literature. *Arch Ital Nefrol Androl* 199062(3):317-22.
4. Gonzglez. Crussi F, Kidd FM, Hernandze RJ. Cystic Nephroma: morphologic spectrum and implications. *Urology* 1982;20(1):88-93.
5. Gallo GE, PENCHANSKY L. Cystic Nephroma. *Cancer* 1977;39(3):1322-7.
6. Heindryckx E, Casselman J., Defloor E, Delanote G. Benign multilocular cystic Nephroma. *J. Belge Radiol.* 1997;80(2)65-7.

