Cystic nephroma in childhood

Sir,
Cystic nephroma is an un-common, benign renal tumour of unknown etiology. Origin is designated as being dysplastic or hamartomous or neoplastic? Two peaks of incidence are seen for this disease.

A 9-month-old female 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 was unremarkable. Blood pressure was 82/54 mmHg. Systemic examination of abdomen showed a 12 x 10 cm 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 showed haemoglobin of 11.2 gm%. Blood urea nitrogen and serum creatinine were 7 mg% and 0.5 mg%, respectively. Urine R/M and C/S showed no abnormality. Ultrasonography detected an 11 x 9.2 cm well-defined multiloculated anechoic lesion. Chest X-ray showed no lesions. Computerized tomography (CT) scan confirmed the sonography findings, adding information of lesion being hypo dense containing clear fluid. No enhancement was seen on contrast.

Provisional diagnosis of cystic nephroma was entertained. Exploratory laparotomy confirmed a 10 x 8 x 5 cm huge, smooth surfaced, cystic renal mass arising from upper pole of left kidney [Figure 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.

Gross specimen showed smooth surfaced kidney as a multiloculated cyst filled with clear fluid and no solid components. Septal thickness was 1.2 mm. Cyst size varied from 2-3 cms each and was non-communicating. Small remnant kidney was present at upper pole with dilated pelvi-calyceal system [Figure 2]. Histopathology showed the cyst lined with columnar to flat epithelium.

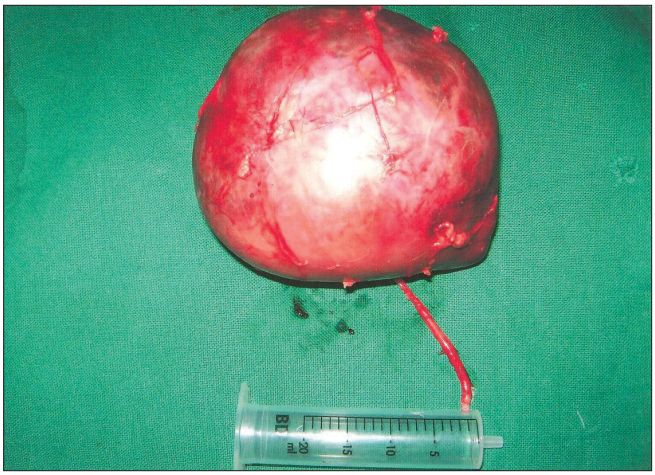


Figure 1: Photograph of gross specimen showing a 15 x 7 x 6 cm huge smooth surfaced cystic renal mass arising from the upper pole of left kidney along with the ureter

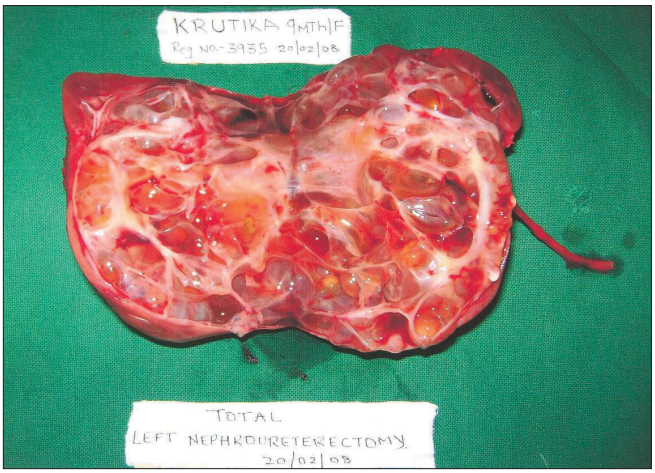


Figure 2: Cut specimen showing multiloculated cysts containing clear fluid. Septae were 1.2 mm thick and non-communicating

Ultra structure displayed cilia. Stroma between the cysts had fibroblastic tissue with dilated vessels but no metanephric blastema. Findings suggested a final diagnosis of cystic nephroma.

Cystic nephroma is an uncommon benign renal neoplasm. The characteristic feature is structurally similar to cystic partially differentiated nephroblastoma.[1,2] The aetiology 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 less than 4 years and 30% in 5th and 6th decade.[3]

It is usually solitary and rarely multiple. Surgical excision 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 and non-filtrating lesions. Few cases have been reported in the literature and the surgical treatment of nephrectomy/nephron sparing surgery is debated. Cystic nephroma having close structural resemblance to nephroblastoma supports, the idea of some surgeons of total nephrectomy as the treatment of choice where biopsy result is ambiguous. 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. The differential diagnosis of cystic nephroma includes cystic Wilms' tumour and cystic partially differentiated nephroblastoma.[4-6] Metastasis or local recurrences have not been reported in cases of cystic nephroma.

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