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MULTILOCULAR CYSTIC NEPHROMA: A RARE CYSTIC TUMOR OF THE KIDNEY IN PEDIATRIC AGE GROUP

Pathology

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ABSTRACT

Among the mixed epithelial-stromal tumours of the Kidney Multilocular cystic nephroma is a, rare(1%-2% of all renal tumours) ,non genetic and benign multicystic lesion.This unilateral renal musticystic lesion shows bimodal age distribution i.e.bellow 2 years old and in adults. MCN ,Cystic Partially differentiated nephroblastoma and Cystic Wilms tumors are a spectrum with benign to malignant nature respectively.

KEYWORDS

Multilocular cystic nephroma ,benign, Sonography/CT ,Radical Nephrectomy.

1.INTRODUCTION

Multilocular cystic nephroma a rare ,non-genetic, benign, unilateral multicystic lesion of kidney . MCN is grouped along with Mixed Epithelial-Stromal tumours of the kidney. This rare cystic lesion can be seen in children below 2 years of old and in adults.normally it is associated with abdominal mass which is usually asymptomatic or showing non specific symptoms.

Sonographic/CT findings relate to the size of the locules and to determine wheather it is a non specific complex intrarenal mass or mass with multilocular configuration and discrete the septa.

Fine needle aspiration has got very little diagnostic benefit because representative samples of most cystic septa cannot be taken.

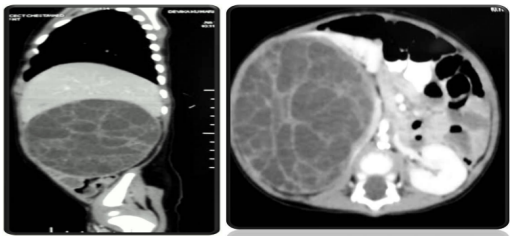
For definite diagnosis Radical Nephrectomy is strongly recommended.

We present an unusual cases of 1 year old female patient with abdominal mass.

2.Case presentation:

An 1 year old girl patient comes with an Abdominal lump with no specific symptom. On examination it is revealed that patient is having lump in the right flank.

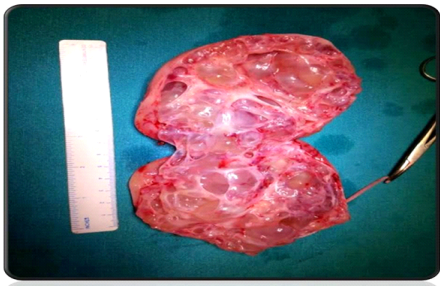
CECT Abdomen shows Multilocular cystic lesion with thin septa replacing entire kidney with no vessel escasement or thrombus formation and no signs of adjacent organ infiltration suggesting provisional diagnosis of multilocular cystic nephroma .



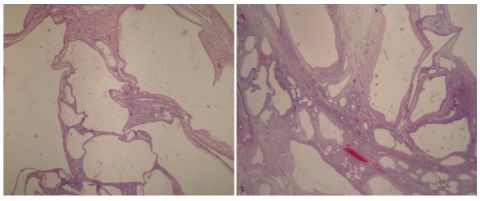
CECT Abdomen:Multicystic lesion with thin septa replacing right kidney.

TREATMENT:As radical nephrectomy is highly recommended in this patient also NEPHRO-UTERECTOMY was performed.During operation it was seen that the tumour had replaced entire kidney with well defined planes with surrounding structures.

Cut section of the affected kidney shows the kidney is replaced with multilocular cystic lesion with thin septa.



CUT SECTION : Multilocular lesion replacing entire kidney.
MICROSCOPIC FEATURES:Histopathologic features shows the lesion is of multicystic nature and cysts are of variable sizes and doesnot contain any immature nephrogenic elements or any solid areas or cytologic atypias. The cystic septas are fibrous with presence of variable amount of inflammation and hypocellular in nature.The lining epithelium is flattened to cuboidal and hobnailing is seen frequently.No ovarian type of stroma is seen .No evidence of any epithelial complexity like branching glands or papillary projections or cribiform structure is seen. Partial or complete fibrous pseudocapsules containing entrapped tubules and glomeruli is common. Some portion of the tumour is seen to be intermingled with normal parenchyma



Patient fully recovered after the surgery and was asymptomatic after the treatment.

3. CONCLUSION

In children who present with cystic renal masses high amount of suspicion should be raised for benign lesions like MCN irrespective of the ages. Pre-operative imaging for MCN is distinctive and can often suggest the diagnosis .Histopathological examination of the operative specimen confirms the diagnosis and diffentiate it from other variants like cystic partially diffentiated nephroma and cystic Wills tumour. The prognosis of patient with Nephrectomy is very good.

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