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CASE REPORT

A RARE CASE OF ADULT CYSTIC NEPHROMA

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ABSTRACT:

50-year-old female , a known case of carcinoma breast was incidentally detected to have a renal cyst in abdominal ultrasonography [USG] done for metastatic workup. Over the span of a year, the cyst gradually increased in size and showed variation in morphology. Laparoscopic left renal cyst deroofing was done. Microscopy and immunohistochemistry (IHC) pointed towards the diagnosis of Adult multicystic nephroma. Here we report a case of rare benign cystic neoplasm of the kidney.

Keywords- renal cyst,ovarian stroma,MEST, meSh terms-renal cyst,ovarian stroma,MEST

INTRODUCTION

Adult cystic nephroma was first described as cystic adenoma of the kidney by Edmunds in 1892 (1). It is a benign cystic lesion of kidneys with a bimodal age distribution occurring predominantly in boys less than 2 years of age and in perimenopausal women of 4th and 5th decades. Incidence accounts for 1 to 2 percentage of all renal tumors and is extremely rare in adults. They usually present with nonspecific symptoms and are often incidentally detected during the radiological examination. Histopathological examination and Immunohistochemical studies are essential to differentiate it from other benign and malignant cystic renal neoplasms. Adult cystic nephroma is composed of multiple cysts separated by fibrous septa with areas resembling ovarian stroma. Surgical excision is the treatment of choice. This is a case of an incidentally detected renal cyst in a 50-year-old female patient with carcinoma breast during metastatic workup.

Case details

A 50-year-old female whose USG performed during workup for carcinoma breast revealed the presence of a cystic lesion with thick septations and a few solid areas which are localized in the upper pole of the left kidney (Figure1).Per abdominal examination revealed a soft firm mass in the left upper quadrant .Computed tomography

(CT) of the abdomen displayed a large exophytic cortical cyst with fine septations measuring 9.5 x 8.5 cm (Bosniak type II F) .No lymphadenopathy or any other lesions were noted. Radiologically the lesion was benign , hence no intervention was done and patient continued treatment for breast carcinoma . Mastectomy followed by chemo-radiation was done . CT scan repeated after one year from the initial scan revealed an increase in the size and change in morphology of the cyst. The size of the cyst was increased to 14.5 x7.8x10.4 cm involving the upper and middle pole of left kidney. Morphologically the cyst wall and septae showed enhancement (Bosniak type III) (Figure 2) . The Pelvicalyceal system and descending colon were seen to be displaced by the cyst. Blood and urine analysis reports were normal. Because of the increasing size and morphological change laparoscopic deroofing of cyst was done and send for histopathological examination. The specimen was received as multiple fragments. On examination few small cysts of size 0.5 cm were seen filled with serous fluid (Figure 3). Microscopic examination revealed multiple cysts lined by flattened to low cuboidal epithelium with bland nuclear chromatin. A few cells showed hobnailing. Cystic spaces were separated by fibrous stroma (Figure 4) . Since this type of morphology can be seen both in Adult cystic nephroma and Cystic renal cell carcinoma IHC markers were done for differentiation. IHC panel (Figure 5) revealed epithelial cell positivity for EMA and PAN CK. Stromal cells showed ER positivity . Both epithelial and stromal cells were positive for Vimentin . CD10 was equivocal . Desmin and AMACR were negative. Hence a diagnosis of Adult multicystic nephroma was given. Postoperative events were unremarkable and the patient is under follow up.

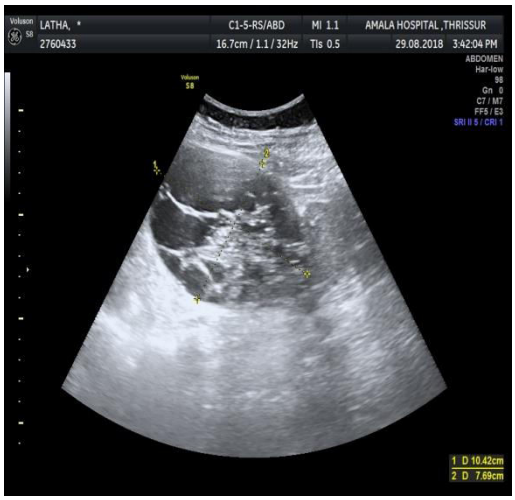

Fig 1 : USG left kidney : Cystic lesion with thick septations and few solid areas in upper pole .

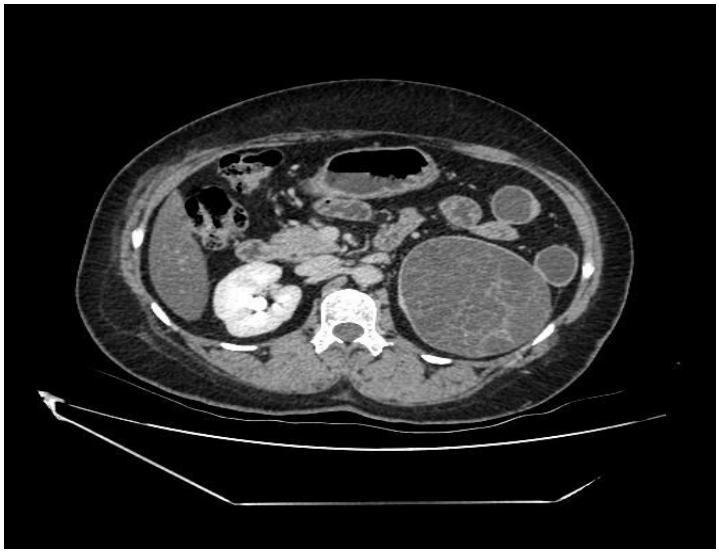

Fig 2 CT left kidney : Exophytic cortical cyst with fine septations measuring 9.5 x 8.5 cm .

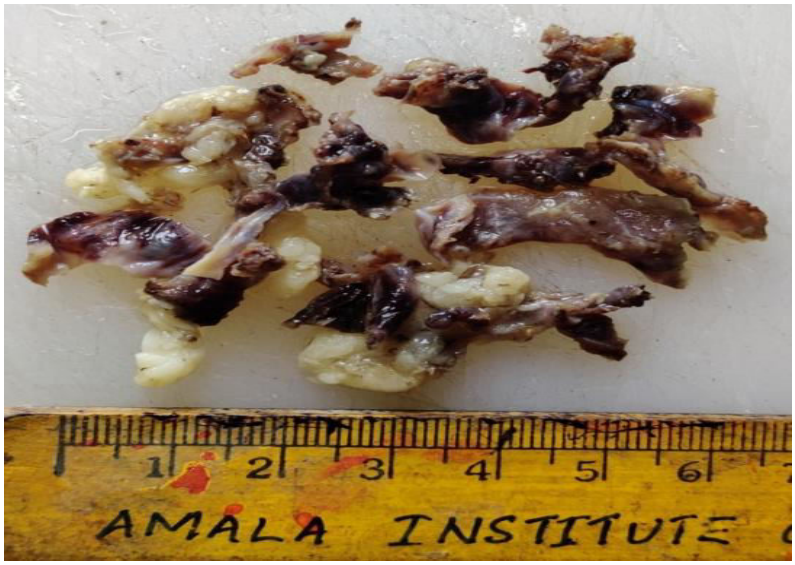

Fig 3: Small cysts of sizes 0.5 cm ,filled with serous fluid .

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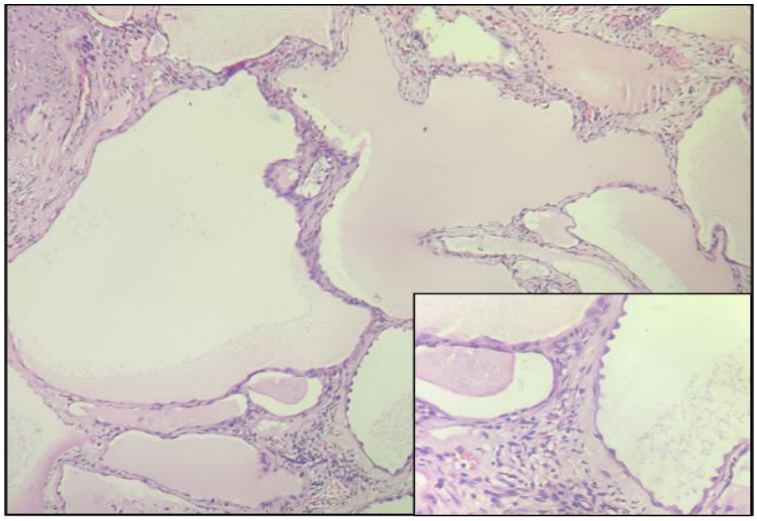

Fig 4: Multiple cysts separated by fibrous stroma and lined by flattened to low cuboidal epithelium with bland nuclear chromatin. Hobnailing present.

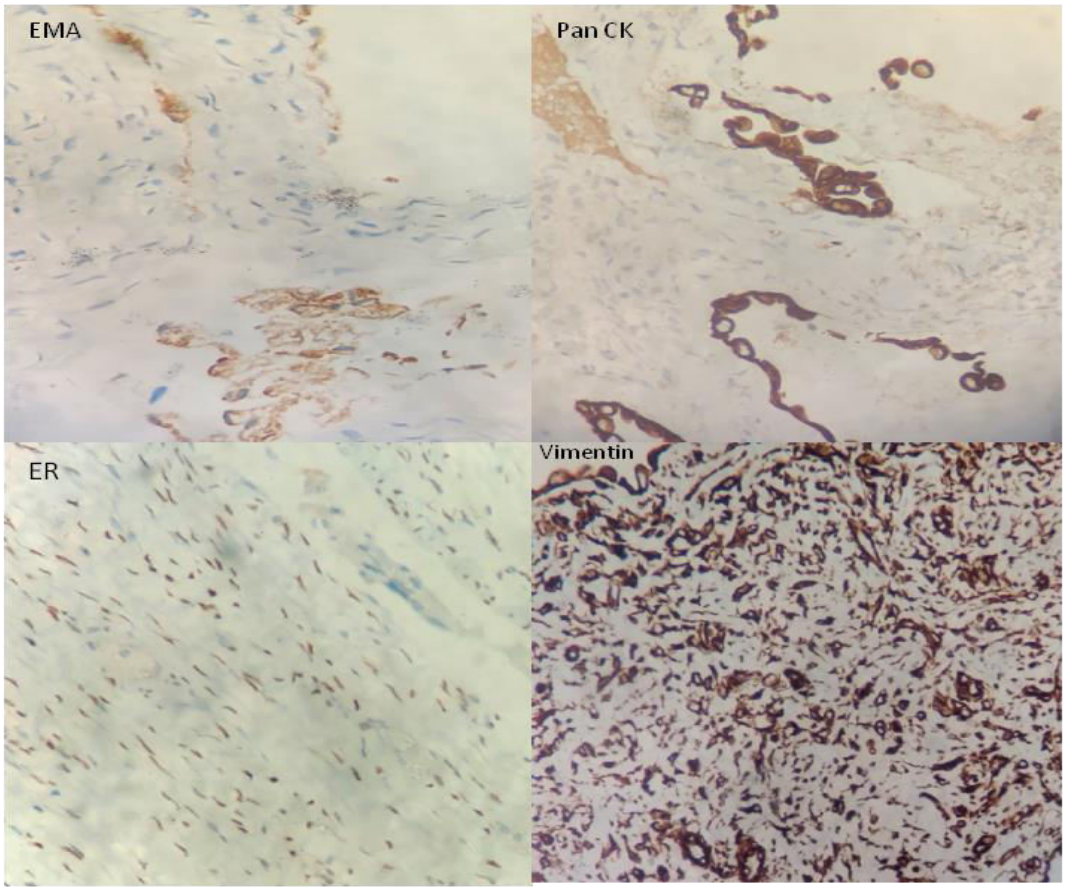

Fig 5: Epithelial cell positivity for EMA and PAN CK. Stromal cells positivity for ER Both positive for Vimentin ..

DISCUSSION

Adult cystic nephroma is classified under mixed epithelial-stromal tumor (MEST) of the kidney as per the "WHO classification of the renal neoplasm's 2016". This neoplasia has a bimodal age distribution with a congenital form occuring in childhood before 2 years of age and an acquired form occurring in adulthood within the 4th and 5th decades of life . Congenital form has a predilection for males and is related to Wilms tumour while acquired form has got a predilection for females (2). The etiology of congenital and adult cystic nephroma is still unresolved, and the classification is controversial. A few theories assert that they arise from ureteric buds as developmental defects(3). Hormonal mechanisms have also been included in the pathogenesis explaining the relative increase in incidence seen in the female population and males who have received hormone manipulation for prostatic cancer (2). Usually, symptoms are nonspecific in adults such as abdominal pain or flank pain, urinary tract infection symptoms, hematuria, and sometimes hypertension. In children they present as palpable abdominal mass . They are often incidentally detected during the radiological examination. The imaging techniques can not differentiate benign cystic nephroma from other benign or malignant complex cystic renal lesions(4). The differential diagnosis considered in children include cystic partially differentiated nephroblastomas [CPDN] and cystic Wilms tumors with minimal invasion . In adults, Clear cell renal cell carcinoma [CCRC] , Multilocular cystic renal neoplasm of low malignant potential ,Tubulocystic renal cell carcinoma and sometimes renal hamartoma can present as cystic lesions with similar clinical and imaging features (5) . Hence histopathological examination with immunohistochemical correlation is essential to reach the correct diagnosis as the growth and prognosis vary considerably between each entity. Diagnostic criteria formed by Eble and Bonsib include an expansible mass composed of cysts and septa encircled by a fibrous pseudo capsule. The interior is lined by flattened, cuboidal, or hobnail epithelium with no expansible solid nodules. The septal cells should resemble mature renal tubules and devoid of the epithelium with clear cytoplasm and skeletal muscle fibers (6). Malignant transformation can occur with sarcomatous or rhabdoid features. IHC studies usually reveal stromal cells positivitivity for CD10, estrogen, and progesterone receptors .The luteinized ovarian like stroma express calretinin and inhibin. Epithelial cells are positive for cytokeratin. Tubulocystic renal cell carcinoma shows clear cell proliferation in the cyst wall and septa and is positive for CD 10 and AMACR. In hamartoma , the tubular epithelium shows positivity for CAM 5.2, epithelial membrane antigen[EMA] , Carcinoembryonic antigen [CEA] and negative for vimentin. CPDN histologically reveals a blastema and/or nephroblastomatous elements. Treatment of choice is surgery which includes excision of the lesion, nephron sparing surgery if the lesion is located away from the collecting system and vascular structures and radical nephrectomy when in doubt of malignancy risk (7). Though there is no risk of metastasis long-term follow-up is essential to rule out local recurrence.(8)

CONCLUSION

Cystic renal tumors are prone to cause confusion in preoperative diagnosis as they have similar clinical,radiological and macroscopic appearances. A definite diagnosis can only

be rendered after surgeryand histopathological examination . Confirmation with IHC is essential to rule out the differentials as treatment modalities vary.

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ABBREVIATIONS

CEA Carcinoembryonic antigen;CCRC Clear cell renal cell carcinoma;CPDN Cystic Partially Differentiated Nephroblastomas ;CT Computed tomography; EMA Epithelial membrane antigen; IHC Immunohistochemistry;MEST Mixed Epithelial Stromal Tumour; USG Ultrasonography

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