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UNILATERAL CONGENITAL MULTICYSTIC DYSPLASTIC KIDNEY MIMICKING CYSTIC RENAL CELL CARCINOMA IN AN ADULT NIGERIAN

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INTRODUCTION

Unilateral congenital multicystic kidney disorder (MCDK) is a developmental disorder in which multiple cysts of varying sizes form in one or both kidneys. It can pose a significant diagnostic dilemma to the clinician when it presents in adults[1]. Cystic tumors on CT scan may be multilocular cystic nephroma, mesoblastic nephroma, cystic renal cell carcinoma, multicystic dysplastic kidney or segmental multicystic dysplasia in a duplicated collecting system[2]. Despite significant advances in diagnostic imaging, the differentiation between these lesions remains difficult[3].

We herein report the challenges posed to us by a case of MCDK.

CASE REPORT

A 58-year old woman presented with a three-month history of recurrent dull aching right loin pain and progressive right loin swelling. Physical examination revealed a middle-aged woman with moderate grade hypertension and a cystic right renal mass.

Abdominal ultrasound (Fig.1) revealed an enlarged right kidney with multiple sonolucent cysts. On contrast-enhanced abdominal CT scan (Fig.2) an enlarged right kidney, hypovascular with multiple septated cysts, a minimal solid component showing as septation and complete flattening of the renal parenchyma was seen. Using the Bosniak criteria, it could be classified as a Bosniak category-3 renal cyst.[3] The left kidney was normal. Serum creatinine, urea and electrolytes were within normal range

A preoperative diagnosis of multilocular cystic nephroma was made, with complex

Fig. 1: Abdominal ultrasound showing multiple cysts in the right kidney

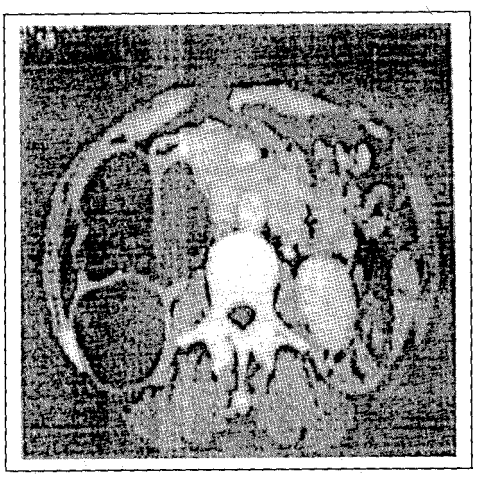

Fig. 2: CT scan of the abdomen showing a multiseptated right kidney and a normal left kidney

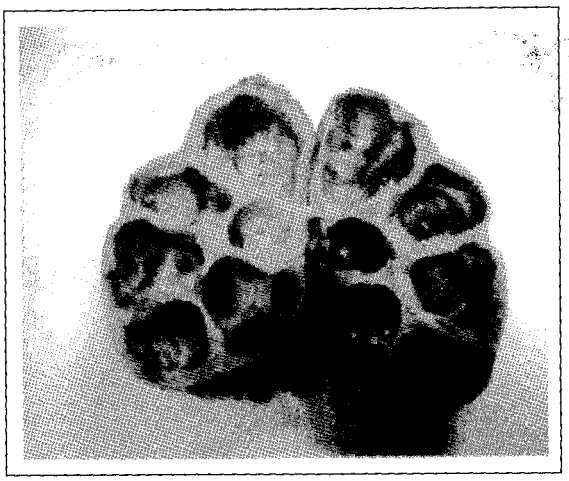

Fig. 3: Right nephrectomy specimen, bisected, showing multiseptated cyst cavities.

renal cyst and multicystic renal dysplasia as differential diagnosis.

The patient was subjected to a right nephrectomy. The operative findings were essentially an enlarged, multiloculated right kidney.

Histopathological examination of the nephrectomy specimen (Fig. 3) revealed a multicystic dysplastic kidney with multiple cysts lined by pseudostratified cuboidal cells. The stroma was composed of glomeruloid bodies, immature tubular and gland-like structures as well as fibromuscular elements. There was no evidence of malignancy.

The postoperative course was uneventful. The patient is being followed up in the outpatient clinic and has remained well six months after her discharge from hospital.

DISCUSSION

Our patient presented in her 6th decade with recurrent right loin pain and a large rapidly growing cystic renal mass which on CT scan appeared as a hypovascular multiple cyst with a minimal solid component showing as septation and complete flattening of the renal parenchyma suggesting the diagnosis of multiloculated cystic nephroma, a complex renal cyst or long standing hydronephrosis. Bosniak proposed a classification of renal cysts and cystic lesions of the kidney into four categories in an attempt to define those lesions requiring no further evaluation, those at risk and – in case of malignant lesions – the degree of malignancy[3,4]. According to this classification which is based on the CT scan features of the cysts, categories 1 and 2 represent lesions which do not require surgical treatment, while categories 3 and 4 represent lesions that need to be treated surgically. However, in a study carried out by Wilson et al.[5], most lesions categorized on CT as grades 2 and 3 were found to be malignant on histology which points to some limitations of the Bosniak classification.

Since imaging facilities are unavailable or poorly developed in most parts of the developing world and, where they are available, great care is needed to distinguish a complex renal cyst (as in our case) from multilocular cystic nephroma, MCDK is more likely to be a post-nephrectomy pathological diagnosis in our environment. Expectant management of such cases is not advisable since there is always a certain risk of leaving a renal cell carcinoma in situ. Thus, our findings in this case underline the need to adequately investigate suspected cases of cystic renal cell carcinoma before and after surgery, as benign and surgically amenable diseases as MCDK may be mistaken for the malignant disease.

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