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Case Report

# CYSTIC, MULTILOCULAR METANEPHRIC ADENOMA OF THE KIDNEY: CLINICAL, RADIOLOGIC AND PATHOLOGIC FEATURES – CASE REPORT

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SUMMARY – A case of multilocular, cystic metanephric adenoma in a 34-year-old man is presented to increase awareness among urologists and radiologists of this rare entity. Metanephric adenoma of the kidney is an uncommon benign epithelial tumor that may present at any age. To our knowledge, only few cases of cystic manifestation have been reported. Metanephric adenoma is extremely rare and its diagnosis should be considered in a patient with well circumscribed multilocular renal tumor and paraneoplastic syndromes. Its appearance on sonography, intravenous pyelography and magnetic resonance is described. Distinct pathologic features are also discussed. Radiologic and clinical features of the tumor are nonspecific, and histopathologic examination is essential to establish definitive diagnosis. Its recognition may facilitate nephron-sparing surgery.

Key words: *Kidney neoplasms – pathology; Adenoma – pathology; Adenoma – radiology; Adenoma – surgery; Case report*

## Introduction

Metanephric adenoma (MA) has recently been recognized as a unique renal tumor characterized by an unusual degree of cellular differentiation and maturation. It is included in the class of benign renal epithelial tumors, with distinctive histologic features. To our knowledge, a little more than 80 cases of this rare condition have been reported in the literature so far, but only few illustrated radiologic features resembling our case. It has indolent clinicobiologic behavior despite the commonly seen large size at presentation. It has occasionally been found responsible for paraneoplastic syndromes such as polycythemia and hypercalcemia[1]. The differential diagnosis includes Wilms' tumor, renal adenoma, papillary renal cell carcinoma, cystic renal cell carcinoma and metastatic tumors.

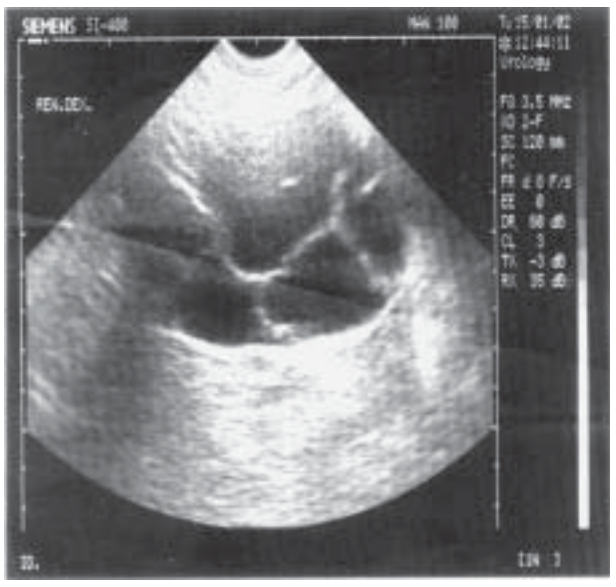


*Fig. 1. Ultrasonography showing a large, well-circumscribed, multilocular, hypoechoic tumor measuring about 10 cm in its largest diameter.*

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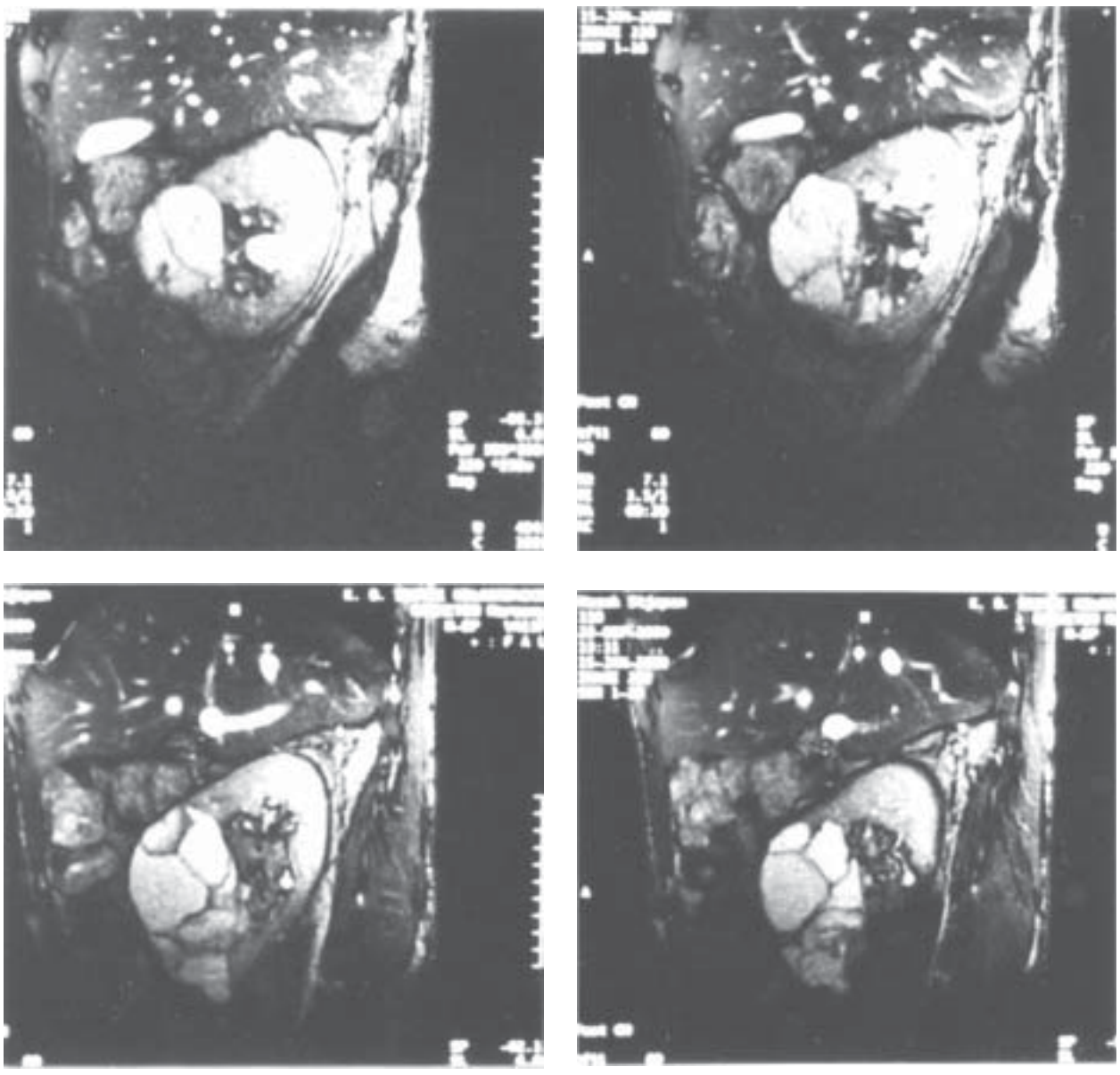

*Fig. 2. Magnetic resonance image showing a multilocular, cystic, hypovascular renal tumor without contrast enhancement.*

## Case Report

A 34-year-old male patient was referred to our department for the evaluation of a mass in the right kidney that had been incidentally detected during an ultrasound examination. His past medical history was unremarkable, except for diabetes. Ultrasonography (USG) disclosed a cystic, multilocular, well circumscribed mass of 10 cm in its largest segment (Fig. 1). To establish the nature and extent of the tumor, intravenous pyelography (IVP) and magnetic resonance imaging (MRI) were employed (Fig. 2). It was mainly consuming the middle and lower portions of the affected kidney, and showed calcifications of irregular multilocular cyst walls. Right kidney USG, IVP and MRI findings suggested a diagnosis of cystic renal cell carcinoma. Based on preoperative radiologic findings and extent of tumor mass, radical right nephrectomy was performed. There was no lymph node involvement. The re-

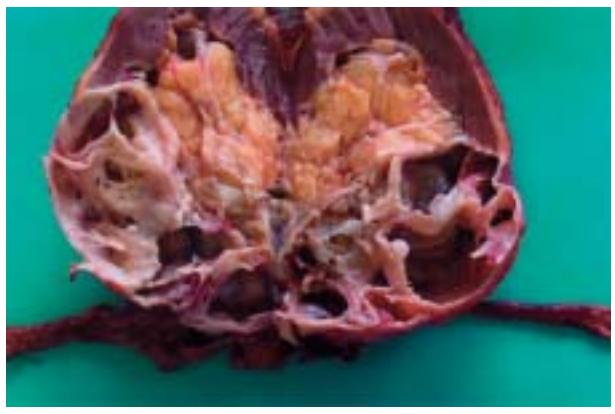

*Fig. 3. Gross resected specimen was well-circumscribed and cystic with solid components, and its cut surface was pale with foci of focal hemorrhage and calcifications of cyst walls consuming a large lower and middle portion of the kidney.*

sected tumor measured 10x10x9 cm (Fig. 3). Pathohistologically, the specimen was composed of small epithelial cells in a tubular or papillary arrangement, without atypia, mitosis, necrosis or invasion of adjacent renal parenchyma or vascular structures, and with abundant psammoma-like calcifications. Occasional tubules, rosettes, and glomeruloid-like structures were seen. Histologic findings were characteristic of MA (Fig. 4). The patient is alive and without recurrence or metastasis 2 years after the surgery.

## Discussion

MA is a kidney tumor first identified in 1988 and classified in the group of nephroblastic tumors[2]. Different terms have been used to describe this entity, such as renal epithelial tumor resembling an immature nephron, nephrogenic nephroma, adult Wilms' tumor, nephroblastoma, or embryonal adenoma. However, MA seems to be the most appropriate term because of its histologic similarity to the metanephric hamartomatous element of nephroblastomatosis[2]. It is described as a benign tumor with no capacity for malignant or metastatic transformation, female predominance at a 2:1 ratio, which can develop at any age. It is generally detected in adults and only occasionally in children. The mean age of these patients is 41 years[2]. The size of MA is highly variable, ranging from 0.3 cm to 15 cm in the reviewed literature[2]. Interestingly, MA is a renal lesion with highest association with polycythemia (12%)[2]. Its chief importance is related to the fact that it is most often misinterpreted as renal cell carcinoma or epithelial Wilms' tumor. To date, only one atypical MA has been reported to metastasize[3]. In most cases, it is incidentally found as a result of an abdominal ultrasound study for signs and symptoms unrelated to the kidney. Less commonly it presents with pain, polycythemia, hematuria, or palpable mass. The key radiologic sign is calcification, which occurs in a statistically higher proportion than in any other renal neoplasia. Also, it is unlikely that MA can be prospectively differentiated from renal cell carcinoma based on imaging features[4]. From the pathoanatomical point of view, these tumors consist of small epithelial cells that form very small acini in an acellular stroma resembling the hamartomatous elements of nephroblastomatosis and Wilms' tumor[5]. Less often, they form tubular, glomeruloid or polypoid and papillary formations. Most also show evidence of regression in the form of scarring and calcification, as in the present case[2]. Rare psammoma bodies are also reported[2]. These lesions are histogenetically related to Wilms' tumor, and are morphologically and immunophenotypically identical to maturing Wilms' tumor and nephrogenic rests[5]. Immunohistochemical and cytogenetic analysis may be helpful in difficult cases. According to its invariably benign course, the metanephric adenoma treatment should be restricted to simple tumorectomy if possible[6]. Despite its size, it is benign and should be distinguished from renal cell carcinoma or Wilms' tumor. Recognition of these tumors is important in rendering the correct diagnosis and assigning a good prognosis for the patient as well as to avoid unnecessary radical nephrectomies.

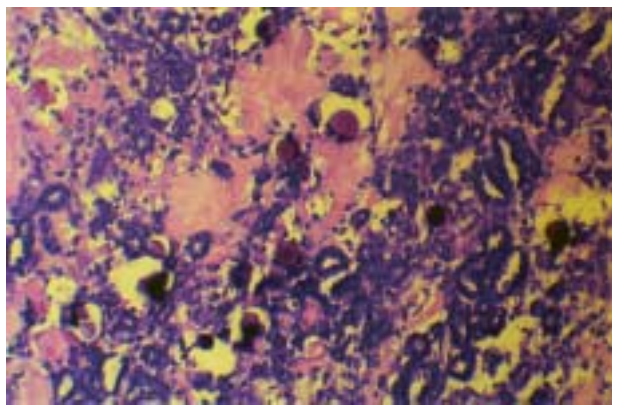

*Fig. 4. Pathologic specimen of metanephric adenoma showing epithelial elements arranged in tubules, short papillae, and glomeruloid bodies with scattered psammoma bodies. No atypia and only rare mitotic activity are present (HE). Immunohistochemically, the tumor cells were reactive for p53.*

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Sažetak

PRIKAZ CISTIČNOG, MULTILOKULARNOG METANEFRIČKOG ADENOMA BUBREGA: KLINIČKE, RADIOLOŠKE I PATOLOŠKE OSOBITOSTI

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Prikazan je slučaj cističnog, multilokularnog metanefričkog adenoma u 34-godišnjeg bolesnika. Metanefrički adenom je izrazito rijetka benigna epitelna neoplazma bubrega koja se može javiti u bilo kojoj dobi. Prema našim saznanjima prikazano je samo nekoliko slučajeva cistične manifestacije metanefričkog adenoma u literaturi. Prikazane su radiološke i patohoistološke značajke tumora. Kliničke značajke tumora su nespecifične i za definitivnu dijagnozu potrebna je patohistološka analiza. Djelomična resekcija odnosno enukleacija tumora terapijska je metoda izbora.


Ključne riječi: *Bubrežne neoplazme – patologija; Adenom – patologija; Adenom – radiologija; Adenom – kirurgija; Prikaz slučaja*