Renal Oncocytosis: Management and Clinical Outcomes

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Purpose: Renal oncocytosis is a rare pathological condition in which renal parenchyma is diffusely involved by numerous oncocytic nodules in addition to showing a spectrum of other oncocytic changes. We describe our experience with renal oncocytosis, focusing on management and outcomes.

Materials and Methods: A total of 20 patients with a final pathological diagnosis of renal oncocytosis from July 1995 through June 2009 were included in the analysis. Patient demographics, intraoperative variables and postoperative outcomes are reported.

Results: Median age at nephrectomy was 71 years (IQR 59–75). Of the patients 15 (75%) had bilateral disease. There were 23 operations (9 right side, 14 left side) performed on 20 patients, and of these procedures 13 (57%) were partial nephrectomies and 10 (43%) were radical nephrectomies. Median dominant tumor mass diameter was 4.1 cm (IQR 3–6.4, range 1 to 14.6). The most common dominant tumor histology was hybrid tumor between oncocytoma and chromophobe renal cell carcinoma in 13 of 23 specimens (57%), followed by chromophobe renal cell carcinoma in 6 (26%), oncocytoma in 3 (13%) and conventional renal cell carcinoma in 1 (4%). Ten patients (50%) had preexisting chronic kidney disease before nephrectomy and chronic kidney disease developed in 5 more after surgery. After a median followup of 35 months no patients had metastatic disease.

Conclusions: Patients with renal oncocytosis usually present with multiple and bilateral renal nodules. Half of the patients had chronic kidney disease at diagnosis and 25% had new onset of chronic kidney disease. No patient had distant metastatic disease during followup. Our management approach is to perform partial nephrectomy when possible and then use careful surveillance of the remaining renal masses.

Key Words: kidney neoplasms; carcinoma, renal cell; nephrectomy

Abbreviations and Acronyms

BHD = Birt-Hogg-Dube
CKD = chronic kidney disease
CT = computerized tomography
eGFR = estimated glomerular filtration rate
GFR = glomerular filtration rate
HD = hemodialysis
NED = no evidence of disease
PN = partial nephrectomy
RCC = renal cell carcinoma
RN = radical nephrectomy

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RENAL oncocytosis is a rare pathological entity in which renal parenchyma is replaced by numerous oncocytic tumor nodules. The disease was first described in 1982 as oncocytomatosis in a case in which both kidneys had diffuse involvement by more than 200 oncocytomas.[1] A subsequent analysis by Tickoo et al showed that each kidney in patients with the disease contained numerous oncocytic nodules in addition to a dominant mass.[2] These nodules usually have a wide spectrum of oncocytic changes rather than the presence of oncocytomas only. The dominant mass typically resembles an oncocytoma, and less often it is a chromophobe renal cell carcinoma or hybrid tumor possessing both histological elements.[2] In addition, the kidneys show a spectrum of other oncocytic changes including oncocytic

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cysts, dispersed oncocytic cells infiltrating between benign nephrons and oncocytic changes in nonneoplastic tubules. Therefore, the authors suggested the more encompassing term of renal oncocytosis.

Some of these features have also been described in patients with the Birt-Hogg-Dube syndrome, an autosomal dominant inherited syndrome with the gene locus located in the short arm of chromosome 17 (17p11.2).[3,4] Individuals with BHD are predisposed to the development of fibrofolliculomas, renal tumors, lung cysts and spontaneous pneumothoraces.[5] In contrast to other renal cancer syndromes patients with BHD present with various renal tumor histological subtypes including hybrid oncocytic tumors, chromophobe RCC and clear cell RCC. More than half of these patients also show features of renal oncocytosis.[6,7]

Approximately 25 cases of renal oncocytosis not associated with the BHD syndrome have been published, most of them as isolated case reports.[1,2,8–15] Our group previously reported on the morphological features and imaging characteristics of renal oncocytosis.[2,16] However, the prognosis and clinical behavior of these tumors remain unclear. In the present study we review our experience treating renal oncocytosis, focusing on the management of the renal tumors and clinical outcomes.

MATERIALS AND METHODS

After receiving institutional review board approval we searched the prospective renal surgery database at Memorial Sloan-Kettering Cancer Center and identified 2,976 patients who had undergone nephrectomy between July 1995 and June 2009. Of these patients 20 had a final pathological diagnosis of renal oncocytosis and were included in the study. All patients had surgical excision of the dominant tumor.

Patient demographic data were collected, and relevant operative and radiographic variables were reviewed. Variables collected from the database included age, gender, clinical presentation, laterality, procedure performed, dominant tumor histology, tumor size, preoperative and postoperative serum creatinine, and followup procedures. For the purpose of the study all specimens were reviewed by a single genitourinary pathologist (SKT). The tumors were classified as chromophobe RCC, oncocytoma or hybrid tumors based on previously published criteria.[2] Hybrid tumors had features of oncocytoma and chromophobe RCC, or tumors with features intermediate between these 2 histological subtypes, and were considered low grade oncocytic RCC, unclassified.

Followup visits were tailored according to final pathology, size of the tumors on the contralateral kidney and renal function. Serial followup imaging was obtained on all patients using CT, magnetic resonance imaging or ultrasound as indicated. Estimated glomerular filtration rate was calculated according to the abbreviated Modification of Diet in Renal Disease equation, accounting for patient age, race and serum creatinine.[17] An estimated GFR of less than 60 ml/minute/1.73 m^2 was considered chronic kidney disease. To assess postoperative renal function, and evaluate the effect of the nephrectomy and disease on renal function, we calculated the GFR for 2 different times, at 1 month after operation and at the last followup.

RESULTS

Patient Characteristics

Median age at nephrectomy was 71 years (range 47 to 82) and 13 patients (65%) were male. At diagnosis 17 (85%) patients were asymptomatic with multiple renal tumors detected incidentally on radiological imaging (figs. 1 and 2). The remaining 3 patients were symptomatic, 2 with gross hematuria, and 1 with weight loss and anemia. Bilateral renal masses

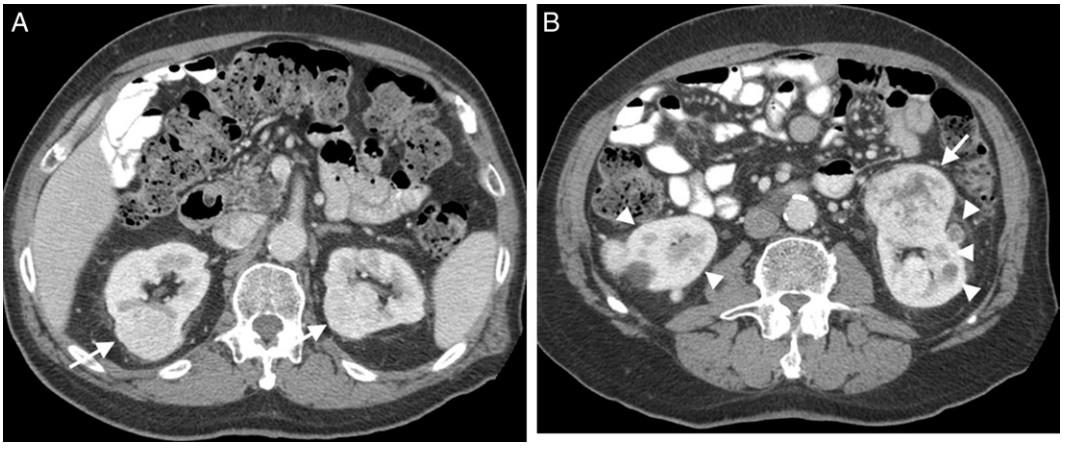

Figure 1. Male patient, 73 years old, with incidental diagnosis of bilateral renal masses. Axial contrast enhanced CT through mid kidneys showed numerous bilateral solid enhancing renal masses (*A*). Axial contrast enhanced CT revealed dominant mass in posterior lower pole of left kidney measuring up to 4 cm (*B*, arrow). Lesion diagnosed as chromophobe RCC on surgical pathology.