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Reappraisal of Morphologic Differences Between Renal Medullary Carcinoma, Collecting Duct Carcinoma, and Fumarate Hydratase-deficient Renal Cell Carcinoma

Chisato Ohe;Steven Smith;Deepika Sirohi;Mukul Divatia;Mariza de Peralta-Venturina;Gladell Paner;Abbas Agaimy;Mitul Amin;Pedram Argani;Ying-Bei Chen;Liang Cheng;Maurizio Colecchia;Eva Comp erat;Isabela Werneck da Cunha;Jonathan Epstein;Anthony Gill;Ondr ej Hes;Michelle Hirsch;Wolfram Jochum;Lakshmi Kunju;Fiona Maclean;Cristina Magi-Galluzzi;Jesse McKenney;Rohit Mehra;Gabriella Nesi;Adeboye Osunkoya;Maria Picken;Priya Rao;Victor Reuter;Paulo de Oliveira Salles;Luciana Schultz;Satish Tickoo;Scott Tomlins;Kiril Trpkov;Mahul Amin;

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Abstract

Renal medullary carcinomas (RMCs) and collecting duct carcinomas (CDCs) are rare subsets of lethal high-stage, high-grade distal nephron-related adenocarcinomas with a predilection for the renal medullary region. Recent findings have established an emerging group of fumarate hydratase (FH)-deficient tumors related to hereditary leiomyomatosis and renal cell carcinoma (HLRCC-RCCs) syndrome within this morphologic spectrum. Recently developed, reliable ancillary testing has enabled consistent separation between these tumor types. Here, we present the clinicopathologic features and differences in the morphologic patterns between RMC, CDC, and FH-deficient RCC in consequence of these recent developments. This study included a total of 100 cases classified using contemporary criteria and ancillary tests. Thirty-three RMCs (SMARCB1/INI1-deficient, hemoglobinopathy), 38 CDCs (SMARCB1/INI1-retained), and 29 RCCs defined by the FH-deficient phenotype (FH-/2SC+ or FH±/2SC+ with *FH* mutation, regardless of HLRCC syndromic stigmata/history) were selected. The spectrum of morphologic patterns was critically evaluated, and the differences between the morphologic patterns present in the 3 groups were analyzed statistically. Twenty-five percent of cases initially diagnosed as CDC were reclassified as FH-deficient RCC on the basis of our contemporary diagnostic approach. Among the different overlapping morphologic patterns, sieve-like/cribriform and reticular/yolk sac tumor-like patterns favored RMCs, whereas intracystic papillary and tubulocystic patterns favored FH-deficient RCC. The tubulopapillary pattern favored both CDCs and FH-deficient RCCs, and the multinodular infiltrating papillary pattern favored CDCs. Infiltrating glandular and solid sheets/cords/nested patterns were not statistically different among the 3 groups. Viral inclusion-like macronucleoli, considered as a hallmark of HLRCC-RCCs, were observed significantly more frequently in FH-deficient RCCs. Despite the overlapping morphology found among these clinically aggressive infiltrating high-grade adenocarcinomas of the

kidney, reproducible differences in morphology emerged between these categories after rigorous characterization. Finally, we recommend that definitive diagnosis of CDC should only be made if RMC and FH-deficient RCC are excluded.

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