Biopsy Proven Medullary Sponge Kidney: Clinical Findings, Histopathology, and Role of Osteogenesis in Stone and Plaque Formation


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Medullary sponge kidney (MSK) is associated with recurrent stone formation, but the clinical phenotype is unclear because patients with other disorders may be incorrectly labeled MSK. We studied 12 patients with histologic findings pathognomonic of MSK. All patients had an endoscopically recognizable pattern of papillary malformation, which may be segmental or diffuse. Affected papillae are enlarged and billowy, due to markedly enlarged inner medullary collecting ducts (IMCD), which contain small, mobile ductal stones. Patients had frequent dilation of Bellini ducts, with occasional mineral plugs. Stones may form over white (Randall’s) plaque, but most renal pelvic stones are not attached, and have a similar morphology as ductal stones, which are a mixture of calcium oxalate and apatite. Patients had no abnormalities of urinary acidification or acid excretion; the most frequent metabolic abnormality was idiopathic hypercalciuria. Although both Runx2 and Osterix are expressed in papillae of MSK patients, no mineral deposition was seen at the sites of gene expression, arguing against a role of these genes in this process. Similar studies in idiopathic calcium stone formers showed no expression of these genes at sites of Randall’s plaque. The most likely mechanism for stone formation in MSK appears to be crystallization due to urinary stasis in dilated IMCD with subsequent passage of ductal stones into the renal pelvis where they may serve as nuclei for stone formation.