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Efficacy of Multi-Detector Computed Tomography for the Diagnosis of Medullary Sponge Kidney

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Key Words
Computed tomography scan • Kidney stone • Nephrolithiasis • Urolithiasis

Abstract
Objective: To expand the diagnostic armamentarium for medullary sponge kidney (MSK), we evaluate the use of high-resolution multidetector computed tomography (MDCT) for MSK diagnosis and compare to the standard intravenous urography (IVU). Despite a significant prevalence amongst stone formers, diagnosis of this well described condition has declined. IVU, the gold standard in MSK diagnosis, has largely been replaced by CT, which has previously been shown unable to demonstrate signs of MSK. ***Methods and Materials:*** Patients with known history of MSK based on IVU underwent limited MDCT urogram. Control group patients, without MSK, also had MDCT urograms performed for other clinically indicated conditions. Studies were scored by board-certified radiologists on a 0–2 scale based on the likelihood of MSK. IVU studies, when available, were similarly graded. ***Results:*** MDCT was diagnostic of MSK in 9 out of the 10 patients with known history of MSK. No false positives were present in our series. The one case of MSK not detected on MDCT was graded as a "1" on its respective IVU. Sensitivity and specificity were 90 and 100%, respectively, when compared with IVU. ***Conclusion:*** Concordance with IVU findings, despite a small reduction in sensitivity, indicates MDCT to be a suitable, and more readily available replacement for IVU in the diagnosis of MSK. Copyright © 2017 S. Karger AG, Basel

Introduction

First described in 1939 by Lenarduzzi, medullary sponge kidney (MSK) is a condition manifested by dilation of the distal renal collecting tubules, often associated with nephrocalcinosis and nephrolithiasis [1]. Patients with MSK are often asymptomatic, but may present with mild hematuria or renal colic [2]. Traditionally thought to be sporadic in nature, many now consider MSK a genetic condition often occurring in association with a number of renal and urinary tract abnormalities and malignancies [3–6]. Though its exact prevalence is unknown, approximately 3–20% of stone formers are thought to have MSK [7].

Some have hypothesized that MSK may be related to the disruption of the interface between the ureteric bud and metanephric blastema during fetal development. Recent work suggests mutations in the glial cell derived neurotrophic factor gene may be causative [8, 9]. Genomic studies have led many to theorize a familial inheritance pattern, potentially increasing the actual prevalence above that of prior estimates [8].

Despite its commonality amongst patients in whom frequent imaging of the urinary tract is necessary, diagnosis of this condition has dramatically declined. Intravenous urogram (IVU) is the traditional gold standard diagnostic test for MSK. Pathognomonic signs of linear or papillary striations representing the ectatic collecting ducts of MSK are often described as reminiscent of

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flower bouquets in severe cases [10]. However, computed tomography (CT), has replaced most older imaging modalities, including IVU, for urologic indications [10].

Although conventional CT is superior to IVU as a diagnostic tool with most urologic conditions, it is typically insufficient to establish the diagnosis of MSK. Conventional single slice CT imaging has been typically performed obtaining images that are 2.5–10 mm thick. At this resolution, comparison studies have revealed CT sensitivity to be markedly lower than that of intravenous pyelography [11].

However, recent years have seen the institution of multidetector CT (MDCT), allowing rapid, high resolution imaging with capability of obtaining images with a slice thickness of 0.625 mm or less. Detailed reconstruction of the renal collecting system with MDCT urography equivalent to that of IVU has been demonstrated [12]. Scattered case series have evaluated the use of MDCT in the diagnosis of MSK, however no comparison study with IVU has been performed to evaluate its reliability. In this study we evaluate the diagnostic ability of MDCT for MSK using IVU as the gold standard.

Methods and Materials

After institutional review board approval was obtained patients with prior history of MSK previously diagnosed with IVU and who required new imaging of the genitourinary tract for other reasons underwent limited MDCT urogram.

The standard scan parameters of a CT urogram were altered to evaluate only the kidneys and, thus, limit radiation exposure in study participants. The first phase included 0.625 mm non-contrast axial images through the region of the kidneys. After the intravenous administration of 100 ml of non-ionic iodinated contrast (iopromide 300 mg iodine/ml, Bayer Healthcare Pharmaceuticals, Wayne, NJ) and 200 ml of normal saline, 7 minute delayed 0.625 mm axial images were obtained. A 64 detector MDCT (Volume CT, General Electric Company Milwaukee, WI) was utilized for all examinations. The nephrographic phase of each study was excluded for analysis.

Additional control patients were similarly evaluated. Included patients underwent MDCT for clinically indicated reasons (i.e. hematuria), but lacked a diagnosis of MSK.

Non-contrast and contrast delayed phase images for both the experimental and control groups were made available in a blinded fashion to 2 board certified radiologists with significant experience in imaging of the genitourinary tract. Study radiologists were blinded to patient history and identifiers, as well as interpretation of their colleague. IVUs and CT's were graded on a 0–2 scale for presence and degree of MSK: Grade 0 representing no MSK, grade 1 representing mild/subtle signs of MSK, and grade 2 representing obvious MSK. The patients' IVUs preceded their MDCT in this study by an average of 71 months (range 54–91 months, in the 5 cases where IVUs were available for review).

Table 1. MDCT and IVU grades for study patients with known MSK

Patient	CT grade I	CT grade II	Consensus CT grade	IVU I	IVU II
1	1	0	0	1	1
2	1	1		1	2
3	1	1		1	2
4	1	1		2	2
5	2	2		2	2
6	2	2		×	×
7	2	2		×	×
8	1	1		×	×
9	1	1		×	×
10	2	2		×	×

Reader 1 = I, reader 2 = II, × = study not available for review. Grade 0 = negative for MSK, Grade 1 = mild/subtle MSK, Grade 2 = obvious MSK.

Differences were settled by consensus read and compared with respective IVU, when available.

Results

MDCT was diagnostic of MSK in 9 out of the 10 patients with prior IVU findings positive for MSK. No false positives were present in our series. Only 5 of the 10 IVUs in patients with a history of MSK could be obtained for review. In 4 of these 5 cases where IVUs were available for review, at least 1 reader scored the MDCT lower than the corresponding IVU. In our series the one case of MSK not detected on MDCT, even with a consensus score, was graded as a "1" on its respective IVU. All of the other 19 MDCTs (9 with MSK and 10 control patients) were graded, with regards to MSK, in agreement between the 2 radiologists. Control arm patients were graded as "0" on all respective MDCT evaluations. The sensitivity in our series for detecting MSK by MDCT compared to IVU as the gold standard was 90% and the specificity was 100%. Table 1 summarizes the scores for the MDCTs and IVUs from the 2 readers.

A number of features identified on MDCT were consistent with MSK. These findings included brush-like parallel striations of contrast in the renal papillae, as well as contrast filled cystic and fusiform dilation of the distal collecting tubules. Calculi, often tiny in size, were easily identified, including those that are considered clinically insignificant (fig. 1 and 2).

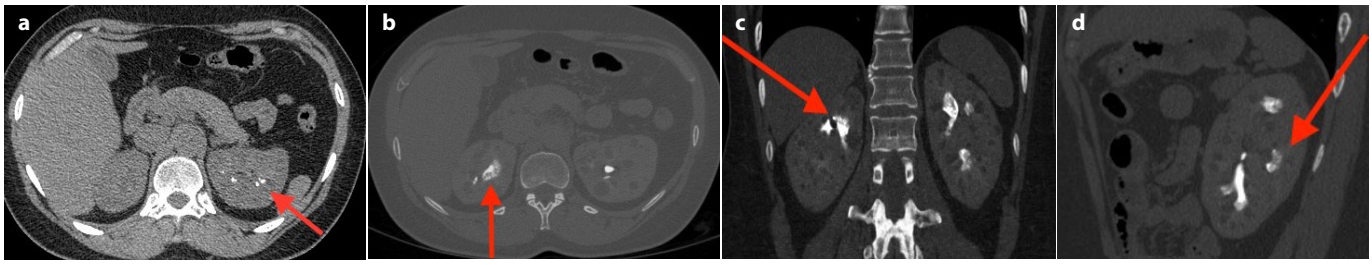

Fig. 1. Pre-contrast axial 0.625 mm source image (**a**), delayed post-contrast axial 0.625 mm source image (**b**) and delayed post-contrast coronal (**c**) and sagittal (of right kidney) (**d**) reformatted images in a patient with MSK. Figure **a** shows medullary nephrocalcinosis involving the left kidney. Delayed post-contrast images (**b**, **c** and **d**) show tubular shaped collections of contrast in the papillary portions of the kidneys which are classic for MSK. This CT urogram was scored a 2 for features of MSK by both readers. The dilated tubules are best seen on the delayed post-contrast images using bone windows (**b** and **d**) as opposed to the soft tissue settings in **c**.

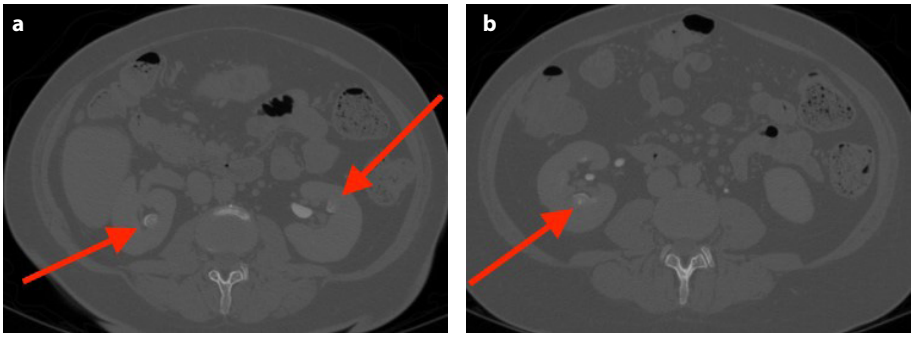

Fig. 2. Delayed post-contrast axial 0.625 mm source images (**a** and **b**) displayed in bone windows in a patient with MSK. These images show more subtle tubular shaped areas of contrast in the renal papillae representing less obvious signs of MSK. This CT urogram was scored a 1 for features of MSK by both readers.

Discussion

MSK is a disease that relies heavily on imaging for diagnosis, with IVU being the traditional gold standard imaging modality. The classic IVU "bouquets of flowers" appearance may only be seen in severe cases. Other pathognomonic signs include brush-like or linear striations, caused by the presence of pooling of injected contrast medium accumulated within the dilated precalyceal collecting ducts [10]. These findings may be localized to a single calyx or extensively involve both renal units, often leading to significant variability in disease presentation [10]. Typical radiographic findings are considered, along with the presence of nephrolithiasis and nephrocalcinosis, in order to diagnose MSK [13].

Although IVU is the gold standard diagnostic instrument, other imaging techniques have been explored. Plain abdominopelvic X-ray can demonstrates nephrocalcinosis of the renal pyramids with clusters of nonobstructive calculi, suggestive but not diagnostic of MSK [14]. Renal ultrasound findings demonstrating medullar hyperechogenicity caused by calculi located within medullary tissue may be seen, but are also nonspecific [14]. Magnetic resonance imaging has a limited role in the imaging of nephrolithiasis, and has shown efficacy in the diagnosis of MSK only in the most severe cases;

T2-weighted imaging can demonstrate significant tubular ectasia [1]. Also, while more invasive, retrograde pyelogram has also been described as an imaging modality capable of diagnosing MSK. It is classically described as a fireworks appearance on consecutively dynamic retrograde pyelography images [16].

Similarly, standard single-slice CT, with or without intravenous contrast, has limited diagnostic accuracy for MSK. Although gross stone burden is often readily evident, standard CT is unable to detect the often scattered distribution and cystic collecting duct dilation unique to MSK [14]. Indeed, prior comparison series revealed significantly lower sensitivity in standard axial CT with 4 mm thick slices versus IVU, with CT's diagnostic capabilities being reliable only in severe cases of MSK [11]. In contrast, within the same study, CT showed superiority in the detection of papillary calcifications. This inability to accurately demonstrate the dilated ductules of MSK on standard single-slice CT is thought to be due to limitations in spatial resolution.

In our series, MDCT urography was utilized, allowing for higher resolution imaging versus conventional CT due to thin sectioning on axial imaging. Koraishy et al. [12] initially showed efficacy of MDCT urography in an analysis of 15 patients with recurrent stone disease. In this non-controlled study, 4 patients were diagnosed with MSK based solely on MDCT, which was able to demonstrate the same pathognomonic findings as IVU with comparable quality. Our findings further confirm the utility of MDCT in the diagnosis of MSK and show excellent concordance with IVU.

Although definitive diagnosis of MSK is not necessary to treat individual stone episodes, radiologic evidence of this condition does have benefits. MSK may be present in as high as 20% of patients with recurrent nephrolithiasis, particularly calcium phosphate or calcium oxalate stones [17]. Although most stones will pass spontaneously, the decline in diagnosis caused by the replacement of IVU with CT for urologic disorders may result in detrimental health outcomes for such patients [10]. Certainly, positive identification with MDCT would allow for closer monitoring, management of the urinary defects common in MSK (i.e. hypercalciuria, hypocitraturia, and alkaline urinary pH), and a thorough discussion of prognosis associated with the condition [10]. Prompt treatment of urinary tract infections, more common in MSK, may lead to the reduction in rates of pyelonephritis and struvite stones along with resultant renal insufficiency or end stage renal disease [18]. In patients necessitating surgical management, documentation of MSK may cause surgeons to preferentially choose ureteroscopy or percutaneous nephrostolithotomy for stones not clearly within the renal collecting system, and, thus, not amenable to extracorporeal shock wave lithotripsy. In severe cases, patients may be counseled regarding the benefit of aggressive therapy such as ileal ureteral replacement or renal autotransplant with pyelovesicostomy to avoid recurrent renal colic episodes. The decline in diagnosis coinciding with limited use of IVU has certainly impacted the utilization of preventative strategies aimed at reducing the morbidity associated with nephrolithiasis.

Furthermore, although initially felt to be sporadic in nature, more contemporary research has revealed familial inheritance patterns, mostly autosomal dominant in nature, which may involve the glial cell derived neurotrophic factor rearranged during transfection interaction [10, 19–23]. Although its molecular basis is incompletely understood, familial screening for MSK may become advisable as its association with congenital disorders (i.e. Beckwith-Wiedemann, congenital hemi-hypertrophy, Wilm's tumor, etc) and prevalence in neonates is further elucidated [10, 24]. MDCT may have a role in the screening of family members of known MSK patients, potentially preventing debilitating stone episodes.

While not as sensitive, for more subtle cases in this small series MDCT appears to provide a suitable replacement for the IVU in the identification of MSK in recurrent stone formers. While prior studies have examined the detection power of MDCT for MSK, this study is the first to utilize a comparison control group and correlate findings with prior IVU. Although not advocating for the replacement of IVU by MDCT, we acknowledge CT as the new mainstay in urologic imaging, and have demonstrated its ability to provide equivalent performance in the diagnosis of MSK. Though some may suggest the diagnosis of MSK is purely academic, it may in fact have far reaching effects ranging from the management of patient prognostic considerations, to guidance of medical and surgical treatment decisions, and providing for further avenues of nephrolithiasis research.

The authors emphasize the necessity of reviewing the thin section MDCT axial source images (0.625 mm or less), utilizing "bone windows" to best evaluate the scans for features of MSK.

There are several limitations to the present study. As this is an initial, proof of concept evaluation, the overall patient numbers are low and limited to one institution. Additionally, several patients in the experimental arm and all control arm patients did not have comparison IVU images available for review. Additional radiation

exposure (MDCT vs. IVU) and cost comparisons were also not performed. Further large, prospective trials are necessary to further determine the role of MDCT in the evaluation of patients with suspected MSK.

Conclusion

MDCT can reliably diagnose MSK. Findings on MDCT may be less striking than on IVU, but it appears to be a reliable first-line imaging modality for establishing the diagnosis of MSK.

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