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Renal manifestations of Kawasaki's disease

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Abstract. Renal sonographic evaluation of seven patients with mucocutaneous lymph node syndrome were performed and correlated with clinical and laboratory data either supporting or not supporting renal disease associated with this entity. Four of seven patients demonstrated significant elevations of the BUN, creatinine and/or significant proteinuria. These four patients had renal sonographic findings of increased cortical echogenicity, enlarged kidneys and enhanced corticomedullary differentiation. This complication of mucocutaneous lymph node syndrome has heretofore not been noted.

Mucocutaneous lymph node syndrome (MCLS) was first described by Kawasaki in 1967 and later reported in the US in 1976 [1, 2]. Other than the mucous membrane and cutaneous manifestations, this disease has been reported to affect the cardiovascular, respiratory and gastrointestinal systems including the gallbladder [3–8]. Renal involvement has not been described other than occasional reports of pyuria, proteinuria and hematuria and one case of hemolytic uremic syndrome [9]. We report seven patients whose kidneys were studied sonographically because of abnormal renal laboratory values reflective of renal disease or because of suspected visceromegaly. This paper discusses the nature of the renal disease, reviews possible pathophysiologic mechanisms and describes the sonographic findings. An illustrative case report will follow.

Case reports (Table 1)

Case 1

A previously healthy 6-year-old black girl developed a sore throat, fever, macular papular rash, cervical lymphadenopathy, vomiting and malaise 2 days prior to admission. The physical examination revealed a temperature of 104° F, lethargy, a macular papular rash over the face, trunk and groin region, erythematous palms and soles, a strawberry tongue, injected pharynx, fissured dry lips and shoddy cervical lymphadenopathy. The neurological and abdominal examinations were unremarkable. A working diagnosis of MCLS or scarlet fever was made.

Admitting laboratory values were: 29000 wbc, hemoglobin 9.9 g, hematocrit 30.7%, blood urea nitrogen (BUN) 67 mg%, creatinine 5.1 mg%, SGOT 109 U/l (normal 7–40 U/l), SGPT 160 U/l (normal 7–4 U/l), total bilirubin 4.1 mg%, negative monospot, negative leptospirosis urine culture, negative throat culture and ASO titers less than 100 todd units. Urinanalysis demonstrated proteinuria, microscopic pyuria and hematuria. Chest X-ray revealed a patchy left lower lobe infiltrate with a normal cardiac silhouette.

On the second day, the urine output decreased and she developed right upper quadrant tenderness. The only significant change on physical examination was a palpable tender right upper quadrant mass. In addition, the creatinine rose to 5.8 mg% and the BUN to 91 mg%. An abdominal radiograph at this time revealed a right upper quadrant soft tissue mass. Ultrasonography confirmed the diagnosis of hydrops of the gallbladder (Fig. 1). Simultaneous examination of the kidneys revealed bilaterally enlarged echogenic kidneys with increased corticomedullary differentiation (Figs. 2 and 3). There was no evidence of hydronephrosis. (Follow up study demonstrated a return to normal).

The patient was carefully monitored for complete renal shutdown. However, over the following week she showed progressive clinical improvement, with BUN, creatinine and bilirubin returning to base line. She subsequently developed perineal and periungual desquamation confirming the diagnosis of MCLS.

Discussion

The clinical picture of MCLS includes fever ranging from 101–104° F for 1–2 weeks, cervical lymphadenopathy, a strawberry tongue, erythema of the oral cavity, dry and fissured lips, bilateral ocular conjunctiva congestion, erythema of the palms and soles and a unique characteristic periungual desquamation at about the 2nd week confirming the diagnosis [1, 2]. Carditis, a frequent complication, accounts for a 1–2% mortality rate.

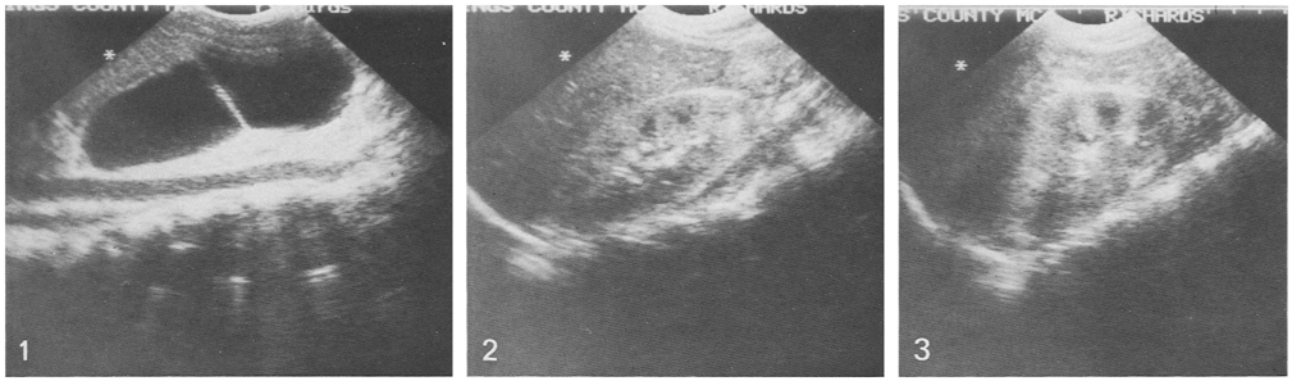

Fig. 1. Longitudinal supine scan of the right upper quadrant reveals a markedly hydropic gallbladder

Fig. 2. The right kidney seen on longitudinal supine scan is diffusely enlarged with increased cortico-medullary differentiation. The renal cortex is more echogenic than the liver

Fig. 3. The left kidney is similarly enlarged with increased echogenicity and prominent pyramids

Table 1. Clinical and sonographic data

Case	Age	BUN/Creatinine (peak)	UA	Sonographic findings
1	16 months	33/0.9	Proteinuria, pyuria, hematuria	Enlarged swollen kidneys, corticomedullary differentiation
2	6 years	91/5.8	Proteinuria, pyuria, hematuria, granular cast	Enlarged swollen kidneys, corticomedullary differentiation
3	6½ years	6/1.2	Proteinuria, pyuria, hematuria	Unilateral enlarged kidney, preservation of the central calyceal complex
4	4 years	54/1.2	Proteinuria, pyuria, Bilrubinuria, hematuria	Enlarged swollen kidneys, corticomedullary differentiation

The etiology continues to remain obscure however rickettsia-like bodies have been implicated with the mite possibly serving as the vector [11–12]. The diagnosis of MCLS is predominantly a clinical one with infectious mononucleosis and scarlet fever completing the differential diagnosis. When complicated by a hydropic gallbladder, the differential diagnosis expands to include leptospirosis, scarlet fever and infantile polyarteritis nodosa. Recovery from MCLS, however, is generally complete.

The vasculitides in this age group include infantile polyarteritis nodosa and Takayasu's disease, both clinically distinguishable from MCLS. However, the pathologic findings of MCLS and infantile polyarteritis nodosa are similar [13]. Renal failure in patients with MCLS has only been noted once in the literature [9]. This case had mild renal failure associated with a microangiopathic hemolytic anemia raising the question of hemolytic uremic syndrome versus renal failure secondary to vascular involvement of the kidneys. However, this distinction cannot be made without renal biopsy.

We feel the sonographic findings of enlarged echogenic kidneys with increased corticomedullary differentiation is best explained by a vasculitis involving the kidneys with resultant fibrinoid deposits and cellular infiltrations (i.e., polymorphonuclear neutrophilic leukocytes, monocytes, lymphocytes, and plasma cells) subsequently leading to ischemia followed by edema [14]. It is speculated that collagen and other similar proteins are deposited and are responsible for the increased echogenicity demonstrated sonographically. However, this cannot be stated for certain as there was no indication for renal biopsy and therefore none was performed.

The mechanism for hydrops of the gallbladder and the intestinal pseudoobstruction seen in this disease is best explained on the same basis as that given here for the renal disease, that is ischemia secondary to vascular insufficiency resulting from a vasculitis [8].

The gastrointestinal and cardiac complications in MCLS have led to a systematic radiological work-up employing several imaging modalities. These include a chest radiograph to evaluate for cardiomegaly and pulmonary infiltrates. An abdominal radiograph to look for a right upper quadrant soft tissue mass when hydrops of the gallbladder is suspected. Sonography however is recommended as the modality of choice since it will show definitively hydrops of the gallbladder without exposure to ionizing radiation [4–7]. Barium studies have been used to evaluate intestinal pseudoobstruction [8]. Angiography has similarly been used to demonstrate vascular complications predominantly seen in the coronary arteries but also the abdominal aorta [3, 10]. Coronary artery aneurysms and occlusions as well as saccular and fusiform aneurysms of the abdominal aorta have been demonstrated.

The seven patients examined in our study had a renal profile ranging from frank renal failure to unremarkable urinalyses. Four of these patients (Table 1) also had sonographically abnormal kidneys compatible with a renal vasculitis [14]. (Three of the latter also had hydropic gallbladders). Of the remaining three, one had trace proteinuria but all had normal BUN/creatinine, and sonographically normal kidneys. However, all four patients with an abnormal renal sonogram had significant proteinuria. The sonographic spectrum of our patients included enlarged echogenic kidneys (four patients), with increased cortico-medullary differentiation. One of the four only had unilateral involvement [15].

Despite the fact that renal failure has not been previously described in MCLS with the exception of one case in association with hemolytic syndrome, we believe that the kidneys can be involved in this disease leading to renal dysfunction ranging from frank renal failure on one extreme to abnormal urinanalyses at the other. The routine urinanalysis and SMA-6 in the seven patients examined sonographically ranged from normal to marked elevation of the BUN and creatinine. However, in our study significant proteinuria (as much as 3+) seems to be a constant finding.

Of the four patients shown to have abnormal kidneys sonographically only three had follow-up repeated sonographic examinations which were considered normal. All four had normal urine laboratory values within a month.

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