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CASE REPORT

Congenital Megacalycosis with IgA Nephropathy: A Case Report and Review of the Literature

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Abstract

Congenital megacalycosis is a rare renal disease characterized by calyceal dilatation without pelvic or ureteral obstruction. If not accompanied by nephrolithiasis and urinary tract infection, this disease is completely benign and does not cause renal dysfunction. We present a case of congenital megacalycosis that was diagnosed at the age of 41 (oldest case in the literature) after admitting with hematuria and acute renal dysfunction. IgA nephropathy was also diagnosed in this patient. Since renal dysfunction is not likely in these patients, if encountered; renal biopsy should be performed although technically difficult to diagnose the cause of this dysfunction.

Keywords: congenital megacalycosis, IgA nephropathy, renal biopsy, renal dysfunction

INTRODUCTION

Congenital megacalycosis is a rare renal disease characterized by calyceal dilatation without renal pelvic or ureteral obstruction. It was first described in 1963.[1] The presentation is mostly unilateral and shows male predominance. It is usually diagnosed in childhood or adolescence.[2] If not accompanied by nephrolithiasis and urinary tract infection, this disease is completely benign and does not cause renal dysfunction.[3] We present a case of congenital megacalycosis that was diagnosed at the age of 41 (oldest case reported in the literature) after admitting with hematuria and acute renal dysfunction.

CASE REPORT

A 41-year-old male patient had been referred to our clinic for acute impairment in kidney function tests. He had a history of bilateral hydronephrosis that was incidentally diagnosed by abdominal ultrasonography 3 years ago. At that time, his renal function and intravenous pyelographic examinations were normal. There was no obstruction or urolitithiasis. He never had a urinary tract infection.

He had experienced a severe upper respiratory tract infection 2 months before and received azithromycin and oseltamivir. Four days later, he complained of dark-colored urine and oliguria. His creatinine level was 2.7 mg/dL. Urinalysis showed hematuria with no protein. Renal ultrasonography revealed bilateral hydronephrosis. The patient was recommended to increase oral intake and was followed for 2 weeks. Although his creatinine level decreased to 1.5 mg/dL, the patient was referred to our center after his hematuria recurred. On admission, the patient's creatinine level was 1.58 mg/dL and estimated glomerular filtration rate (GFR) was 49 mL/min/1.73 m^2. Blood pressure was 130/80 mm Hg and the physical examination was unremarkable. He had dysmorphic erythrocytes on urine sediment without marked proteinuria or eosinophiluria. Sizes and parenchymal thicknesses of both kidneys were normal on ultrasonography. There was mild to intermediate bilateral hydronephrosis in collecting systems with no dilatation in ureters. Magnetic resonance urography revealed

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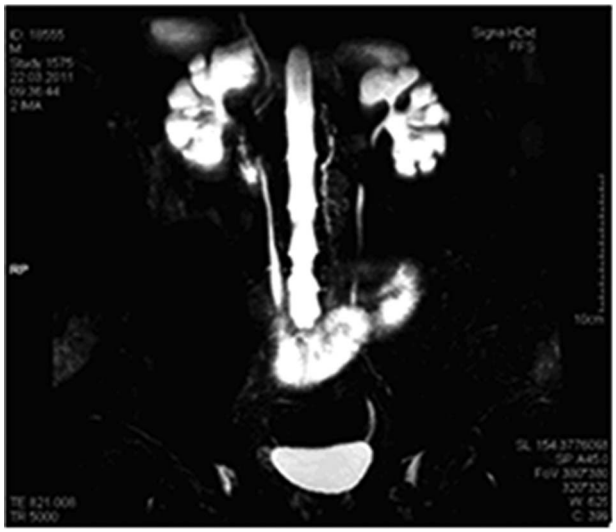

Figure 1. Magnetic resonance urography revealed calyceal dilatation, increased number of calyces, and normal ureters and bladder.

increased number of calyces and renal pelvic dilatation. Ureters and bladder were normal and obstruction was excluded (Figure 1). He was diagnosed with megacalycosis. Renal biopsy was performed to reveal the cause of hematuria and acute kidney dysfunction. Biopsy represented a sample from a small area in renal cortex and included 15 glomeruli, four of which were globally sclerotic. Glomeruli looked normal light microscopically (Figure 2). No mesangial, endocapillary or extracapillary proliferations were present. Necrosis, karyorrhexis, or

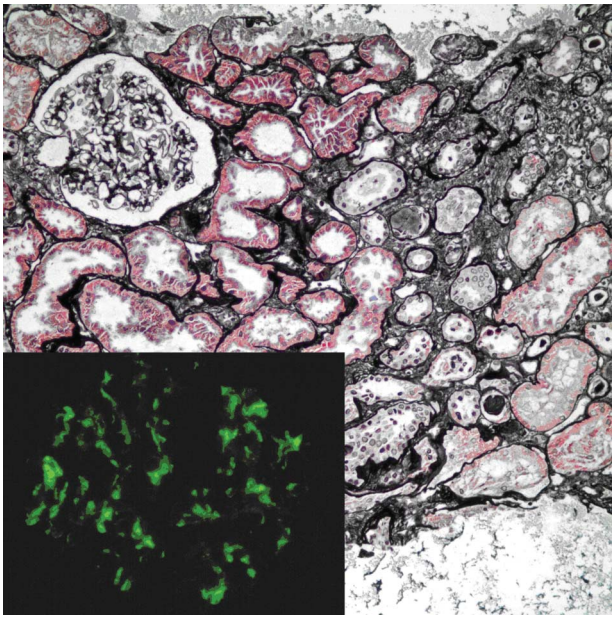

Figure 3. Mild tubular atrophy with interstitial fibrosis is noted at right side of the picture (Jone's methanamine silver stain, × 200 magnification). Inset shows deposition of IgA in the mesangial regions (imunofluorescence, fluorescein isothiocyanate-conjugated anti-IgA antibody ×400).

segmental glomerular sclerosis was not identified. Mesangial regions were not expanded significantly. There were a few narrow foci of mild tubular atrophy and interstitial fibrosis (Figure 3). Interstitial inflammatory infiltrate was not observed except a small amount of

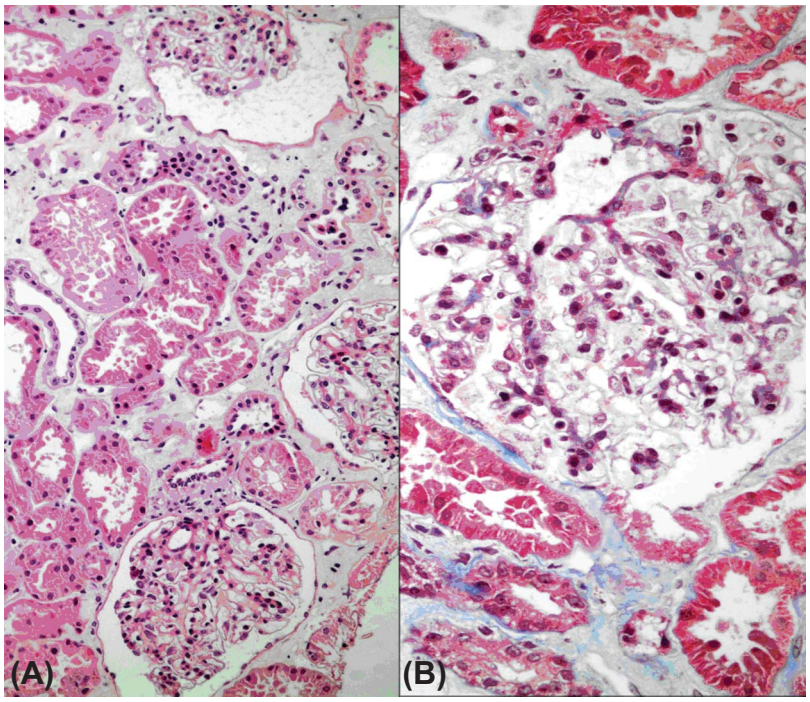

Figure 2. Glomeruli had no significant alterations light microscopically [(A) hematoxylin and eosin stain, ×200 magnification and (B) Masson's trichrome stain, ×400 magnification].

lymphocytes in the subcapsular scar region. Arteries revealed mild to moderate intimal fibrosis. Immunofluorescence microscopy showed diffuse and global IgA deposition in the mesangial regions (Figure 3, inset). C3 accompanied IgA in glomeruli. Staining for IgG, IgM, C4 and C1q was negative. The histopathological diagnosis was IgA nephropathy. Prednisolone (1 mg/kg/day) was started for the patient to treat IgA nephropathy. However, he could not tolerate prednisolone due to side effects, and conservative treatment was recommended with an angiotensin receptor blocker to control blood pressure. Under this treatment, serum creatinine level of the patient was 1.3–1.4 mg/dL.

DISCUSSION

Congenital megacalycosis is a rare congenital abnormality. The renal calyces in affected kidneys are enlarged without any evidence of obstruction in infundibula, pelvis, or ureter.[2,3] In addition to dilatation, the calyces may be increased in number. Renal parenchymal thickness and structure are often normal. This anomaly is encountered predominantly in male Caucasian patients, and usually presents unilaterally.[2]

Congenital megacalycosis is thought to occur due to abnormal development of the renal medulla, which leads to hypoplastic renal pyramids and blunted, dilated calyces.[4] Renal papillae are hypoplastic and, instead of protruding into the calyceal cavity, they surround hypotonic calyces. Urinary stasis in those dilated calyces may predispose to infection and stone formation.[5] Abnormalities such as megaureter or Schinzel–Giedion syndrome may accompany this condition.[3,5–7]

This disorder is generally diagnosed during evaluation of nephrolithiasis and/or urinary tract infection.[7–12] Obstructive nephropathy and vesicoureteral reflux should be excluded before diagnosing congenital megacalycosis. Ultrasonography, intravenous pyelography, scintigraphy, and computed tomography play important roles in the exclusion of obstructive causes.[13,14] We preferred magnetic resonance urography as the diagnostic imaging modality, because of the risk of contrast nephropathy.

Patients described in the literature were mostly children, and the oldest patients were 22 years of age.[7,8] Probably because of asymptomatic course of our patient, he was diagnosed at the fifth decade, and he is the oldest patient in the literature at the time of diagnosis. Clinical characteristics of all the reported patients with congenital megacalycosis were shown in Table 1.[7–12,15–18]

Renal function of patients with congenital megacalycosis is usually normal both at the time of diagnosis and during follow-up.[12] Slight defects in concentration capacity can be seen due to relative loss of juxtamedullary nephrons.[2] There were case reports of patients who had normal renal GFRs, but had parenchymal injury due to recurrent urinary tract infections.[8] If there is renal dysfunction in patients with congenital megacalycosis that cannot be explained by simple blood, urine, and

Table 1. Clinical and demographic characteristics of the cases with congenital megacalycosis described in the literature.

Author(s), year	Age/gender	Calycosis	Clinical presentation
Reyes and Trejo, 1982[15]	Newborn	Bilateral	n/a
Vargas and Lebowitz 1986[7]	Six patients ages between 8 and 22 years three boys and three girls	Unilateral in all patients	Urinary tract infection (4/6), flank pain (1/6), hematuria (1/6) and megaureter in all patients
Shieh et al., 1990[16]	Seven patients ages between 7 and 14 years Five boys and two girls	Three bilateral, four unilateral	Asymptomatic (6/7) and hematuria (1/7)
Parlaktas et al., 2004[10]	20-year-old male	Bilateral	Bilateral lombar pain
Kasap et al., 2005[8]	17-year-old male 4-month-old boy	Bilateral Bilateral	Urinary tract infection Antenatal hydronephrosis
Redman and Neeb, 2005[12]	13-year-old boy	Unilateral	Gross hematuria after trauma
Alivanis et al., 2006[9]	22-year-old male	Bilateral	Urinary tract infection
Kleszczynski et al., 2006[17]	10-year-old girl	Bilateral	Urinary tract infection
Alivanis et al., 2006[9]	22-year-old male	Bilateral	Urinary tract infection
Pieretti et al., 2009[11]	2-year-old boy 9-month-old girl	Bilateral Unilateral	Antenatal hydronephrosis Accidentally in ultrasound
Bekele and Sanchez, 2010[18]	8-year-old boy	Unilateral	Antenatal hydronephrosis

Note: n/a, not available.

radiological tests, a renal biopsy should be performed to diagnose the other causes such as glomerulonephritis. Management of congenital megacalycosis includes prompt treatment of urinary tract infections and nephrolithiasis. Patients should be followed annually by kidney function tests and urinalysis.[12] To minimize the risk of stone formation, liberal fluid intake is advised. Prognosis is very good in these patients. The patient in this report had hematuria and impaired renal functions without an obvious cause such as urinary tract infection or nephrolithiasis. He also had a history of a severe upper respiratory infection requiring use of antibacterial and antiviral medications.

After excluding urinary tract infections and nephrolithiasis, IgA nephropathy is one of the most common causes of hematuria in general population. Approach to renal dysfunction in patients with congenital megacalycosis is not well described. In this case, a renal biopsy was performed to elucidate the cause of hematuria and renal dysfunction. Although renal biopsies performed by experienced physicians have low probability of causing severe complications, dilated calyceal system and relatively thin renal parenchyme make renal biopsy technically challenging in patients with congenital megacalycosis.[19] The result of biopsy in this case was IgA nephropathy, consistent with the clinical picture including hematuria and renal dysfunction. This is the first case of IgA nephropathy accompanying congenital megacalycosis in the literature.

In conclusion, congenital megacalycosis does not cause symptoms if not associated with nephrolithiasis or urinary tract infection and has a benign prognosis. Since renal dysfunction is not likely in these patients, if encountered; renal biopsy should be performed although technically difficult to diagnose the cause of this dysfunction.

***Declaration of interest*:** The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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