

SHORT REPORT

Congenital Megacalycosis: A Rare Renal Developmental Anomaly

Bekir Suha PARLAKTAŞ, Fikret ERDEMİR, Nihat ULUOCAK
Department of Urology, Faculty of Medicine, Gaziosmanpaşa University, Tokat - Turkey

Received: April 15, 2004

Key Words: Megacalyces, megacalycosis, congenital

Congenital megacalycosis (CM) is a rare developmental anomaly of the kidney characterized by nonobstructive dilatation of the renal calyces (1-3). It is thought to occur due to abnormal development of the renal medulla, which leads to hypoplastic renal pyramids and blunted, dilated calyces (4). This nonobstructive dilatation of the calyces causes urinary tract infection (UTI) and calculus formation because of stagnant urine. Patients therefore usually present with symptoms of these coexisting entities and the condition is diagnosed from these symptoms (5,6). If differential diagnosis is made from obstructive conditions inappropriate surgery is avoided and presenting clinical conditions respond well to conservative therapy (1,3). We report a case of bilateral CM and the relevant literature is reviewed.

Case Report

In January 2004 a 20-year-old male was admitted to our clinic with a history of bilateral lomber pain with a preponderance to the left. He had been suffering from this complaint for about 10 years, and a series of urographies had been performed several times during this period. He had been followed up with examinations at irregular intervals and had undergone no surgical intervention. However, he had been given appropriate medical therapy for pain and UTI several times. Physical examination revealed a normal healthy male. Routine hematologic and biochemical tests were normal, than a microscopic hematuria in urinalysis. Culture for urine was negative. An intravenous pyelography IVP update was needed. This was performed, and the findings of this recent urography were as follows: the calyces were

dilated and increased in number in both kidneys. The axes of the kidneys were rotated with normal ureteropelvic junctions. The ureters were not dilated and they seemed to be nonobstructed bilaterally (Figure 1). Ultrasonographic examination showed dilatation of the calyces in both kidneys with a normal pelvis. The kidneys were of normal length and the parenchyma was normal in thickness. In addition, there was a calculus (10 x 8 mm) in the lower pole of the left kidney. Renal scintigraphy with technetium 99m diethylene-triaminepentaacetic acid (Tc 99m DTPA) showed a normal urinary flow and there were no obstruction in the anomalous kidneys. A technetium 99m dimercaptosuccinic acid scan (DMSA) showed a nonobstructed washout pattern in both kidneys with a normal split renal function, and there was no scar in either kidney. The IVP and renal scintigraphy findings were in agreement with the criteria of the disease in the literature, and this led us to a diagnosis of CM. We therefore placed the patient into a follow-up program and performed ESWL for the left lower pole stone. After a lithotripsy trial the kidney was free of stone. At the end of 3rd month the patient was symptom free with no complaints and he was referred for another yearly check up including IVP and routine blood and urine assays.

CM was first described by Puigvert in 1963 (7). The reported cases in the literature have been predominantly males (1-3,6). It is widely regarded as a primary disease of nonobstructive nature. Renal function in the affected kidney is therefore normal and neither functional nor anatomic deterioration generally occur in long term follow-up (1-3,8). The difference between CM and an obstructive uropathy is important because renal function



Figure 1. Intravenous urogram showing dilated calyces in both kidneys. Ureteropelvic junctions are normal without normal non distended pelvis and normal dependent ureters. Axial rotation of the kidneys is also noted.

is gradually compromised in obstructive hydronephrosis (9).

CM is characterized by enlarged kidneys and uniform dilatation of all the calyces. The infundibula, pelvis and ureter are normal and there is no obstruction in the collecting system. In addition to dilatation the calyces maybe increased in number (polycalycosis). Cortical tissue is usually normal in thickness and there is no cortex abnormality such as scarring or signs of chronic infection. The affected kidney is generally larger than normal and renal functional tests show normal results (1,3,4,6,8-10). CM has also been described as an associating feature of a rare syndrome known as Schinzel-Giedion Syndrome (SGS). This syndrome is characterized by multiple congenital malformations, including midfacial retraction, genitourinary and renal malformations, multiple skeletal abnormalities, severe psychomotor impairment, seizures and CM (4). Patients with CM usually present with symptoms of UTI and urinary calculus. These include pain,

hematuria, fever and dysuria (2,5,6). Stasis of urine in enlarged, redundant calyces promotes the formation of urinary calculi and infection (2,6).

In the differential diagnosis of CM some clinical conditions causing obstructive uropathy, intermittent hydronephrosis and vesicoureteral reflux should be considered (1,3,10). IVP shows typical dilatation of the calyces without enlargement of pelvis and supports the diagnosis of CM. Ultrasonography (US) usually shows the same findings. Absence of reflux in voiding, cystourethrograms and a normal wash-out pattern in diuretic renal scintigraphy are further radiologic supporters of the diagnosis of CM (1-4,6-10). Among these modalities IVP gives the best functional picture and anatomic structure of the kidney, which are not provided by the others. With the help of US images and scintigraphic tracing findings, IVP clarifies the clinical picture and plays the most important role in diagnosis (1,10).

Radiographic findings and renal function tests have remained stable in CM cases that were followed up for several years (1,3). This situation is also relevant to our patient, because despite a long period with no surgical intervention there is no difference or deterioration in urographies as far as renal size, parenchyma, calyces, pelvis or ureters are concerned.

Surgery is not necessary for the treatment of a primary anomalous kidney with CM but the presence of a stone and infection mandates appropriate therapy (1,3). Treatment alternatives may be follow-up, ESWL, and sometimes surgery for urinary calculus, and in order to control supervening UTI appropriate antibiotic therapy is administered. To minimize the risk of stone formation more fluid intake is advised (1,5). In the long term follow-up period the patients should be in a continuous and close follow-up program because of the high risk of infection and stone formation, and annual check-up with routine blood chemistry, urinalysis and IVP are recommended (3).

Corresponding author:

Bekir Suha PARLAKTAŞ

Mevlana Sitesi, 2. Blok

Kat. 2, No. 5

60100- Tokat - Turkey

e-mail: bsuha@mynet.com

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