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Differential Diagnosis and Management of Megacalyces (Puigvert's Disease): A Case Report

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ABSTRACT

Megacalyces is a rare congenital urinary anomaly consisting of nonobstructive dilated calyces. The renal pelvis and ureter have a normal caliber. We report an asymptomatic unilateral megacalyces in a 22-year-old woman. She had an increased number of dilated calyces on the right kidney and a cluster of small calculi over the lower pole. The largest stone in the lower calyx was 5 mm. The renal pelvis and ureter were normal and there was no evidence of obstruction. In the absence of functional symptoms and because of the small urinary stone size, we decided on surveillance. A follow-up examination 22 months later showed that the patient is still asymptomatic and the stone sizes are unchanged. We discuss literature related to the clinical features, differential diagnosis, and management of this condition.

KEYWORDS: Malformation; Congenital; Renal abnormalities; Calyx; Megacalices
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Abbreviations and Acronyms
CT, computed tomography
IVU, intravenous urography

INTRODUCTION

Megacalyces is a congenital malformation characterized by a nonobstructive dilatation of all renal calyces [1]. The diameter and morphology of the ureter and renal pelvis are normal [2].

Diagnosis of megacalyces is often fortuitous during intravenous urography (IVU) or renal ultrasound [2]. The disorder is not progressive and it does not require any surgery [1]. We discuss the clinical, radiological, and evolutionary information about this condition through the following case report.

CASE REPORT

The patient was a 22-year-old self-identified Caucasian woman. A gastroenterologist had performed an abdominal ultrasound because of intermittent constipation and incidentally noted a right renal stone.

Her medical history was not significant. There was no history of renal disease, renal colic, infection, hematuria, or stone passage. She was totally asymptomatic and the physical examination was unremarkable. Urine analysis and hematological and biochemical blood tests were normal. Urine cultures for bacteria and acid-fast bacilli were negative.

A plain abdominal (kidney, ureter, bladder) radiograph demonstrated a cluster of small calculi over the lower pole of the right kidney (Figure 1). IVU showed an increased number of dilated calyces on the right kidney and a 5 mm stone in the lower calyx; the renal pelvis and ureter were normal (Figure 2). There was no evidence of obstruction. The normal cortical thickness was obvious in the nephrogram phase. The left kidney was of normal size, with a normal collecting system. A computed tomography (CT) scan confirmed the marked right calyceal dilatation (Figure 3). In technetium-99m-MAG3 renal scintigraphy, the right kidney clearance was 42% of the total renal function with stasis in the upper urinary tract (Figure 4). All of these radiological investigations were compatible with the diagnosis of megacalyces. We did not perform cystoscopic examination to rule out the possibility of reflux to the right kidney or neurogenic bladder because the diagnosis was evident to us.

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case report

Figure 1. Abdominal Radiograph Showing Calculi Over the Lower Pole of the Right Kidney (arrow).
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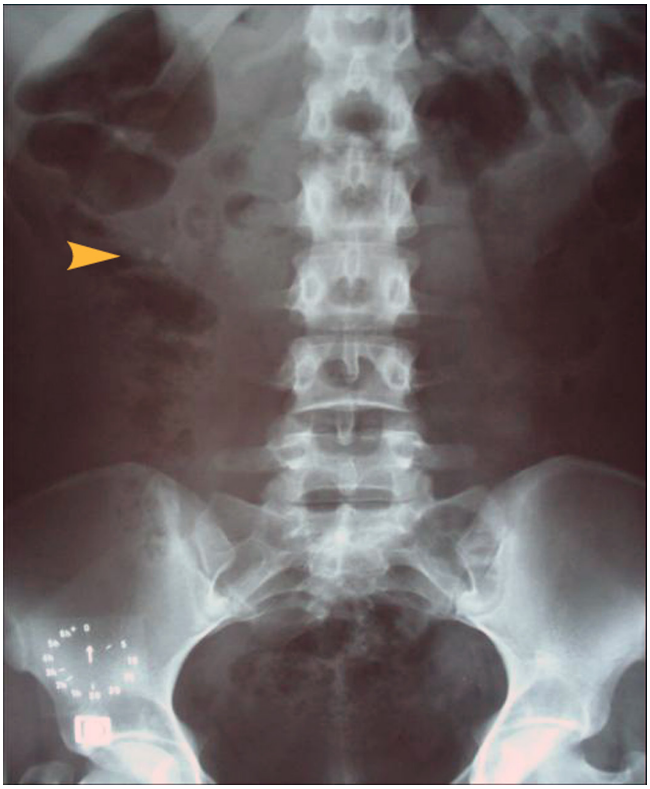

Figure 2. Intravenous Urography Showing Dilated Calyces on the Right Kidney With a Normal Pelvis and Ureteral Caliber.
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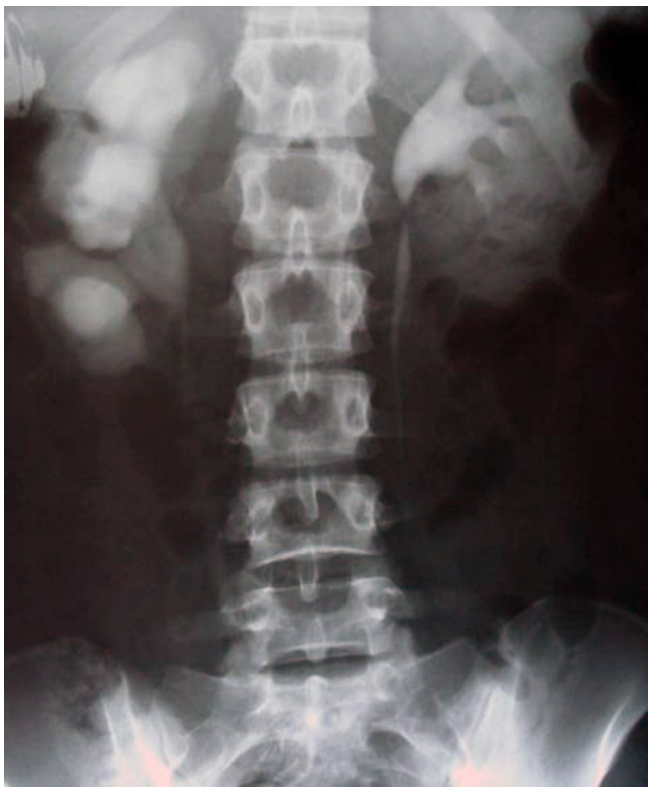

In the absence of functional symptoms and because of the small urinary stone sizes, we decided on surveillance. A follow-up examination 22 months later showed that the patient is still asymptomatic and the stone sizes are the same.

DISCUSSION

Megacalyces, also known as Puigvert's disease, is a clinical entity characterized by homogenous dilatation of all renal calyces and a renal pelvis and ureter of normal caliber [1,3,4]. It was first described Puigvert in 1963 [1]. Congenital megacalyces is a developmental condition of the kidney consisting of underdevelopment of the renal pyramids and nonobstructive dilatation of the calyces [1,2,5,6]. In fact, the single most important feature is the absence of any obstructive element [5,6]. This condition is benign and nonprogressive [7]. In the past, it has been misdiagnosed as obstructive hydronephrosis with unwarranted attempts at surgical therapy [5]. Thus, we report this case and point out diagnostic criteria.

The affected kidney is usually larger than normal and it has

Figure 3. Computed Tomography Scan Showing Enlarged Right Calyces.
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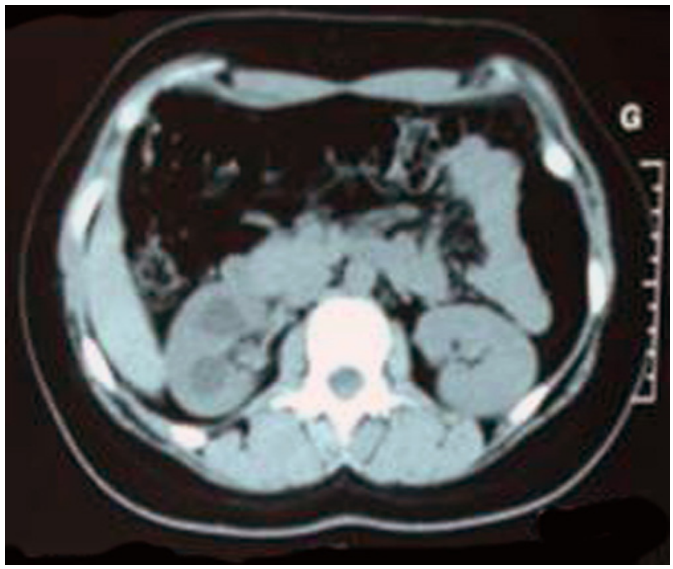

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Figure 4. MAG3 Scintigraphy Showing a Normally Functioning Right Kidney.
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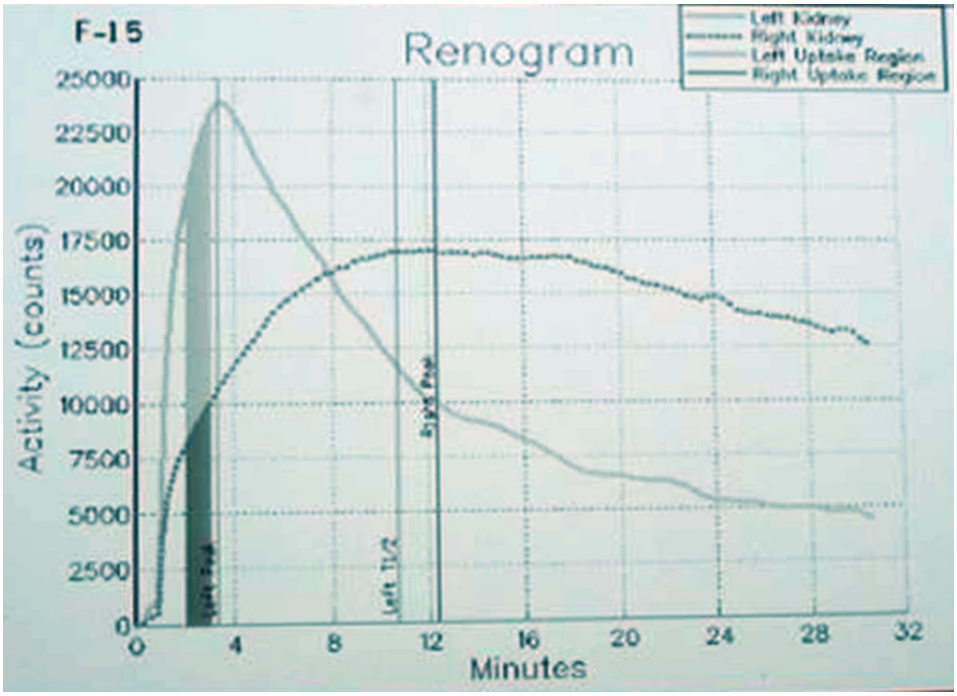

prominent fetal lobations [5,8]. It should be considered as a primary anomaly of the renal pyramids in which the enlarged calyceal cavities take the space of normal renal pyramids [5]. The kidney usually has normal function in noncomplicated cases [8,9]. Histologic examination shows a normal renal cortex and a characteristic arrangement of collecting ducts. The ducts are shorter than normal and oriented transversely, instead of having the usual longitudinal orientation [1,8,9].

Our patient has unilateral megacalyces (the most common condition), but it may be bilateral [5,10]. The patient age at diagnosis may range from 5 to 60 years, but the majority are in the third and fourth decade [5]. It is more common in males than females [5,9,10] and particularly affects people of Caucasian race [11].

Etiology

To date, the pathogenesis of megacalyces remains unclear [3,12]. A congenital defect has been suggested but not proven [5]. One case was diagnosed by prenatal ultrasound and some cases have been reported in the newborn [13]. Puigvert [2] and others [5,14] suggested an embryologic fault during the early divisions of the ureteropelvic anlage or ureteral bud when it is joining with the metanephric blastema. Some observations support the genetic nature of the disease [6,15]. Others blame a discrepancy in the muscular development of the calyceal wall and development of the protuberant papilla in the early stage of intrauterine life [16]. Primary megaureter and megacalyces occasionally coexist [17].

Diagnosis

There is a greater lithogenic and infection (secondary to calyceal stasis) predisposition in patients [3,5]. These complications usually bring the patient to medical attention. However, this entity is often symptom-free. The presence of symptoms should suggest an infection, stones, or a primary obstructive cause [18]. Hematuria [5], flank pain, nausea, and vomiting [6] may be seen. To establish the differential diagnosis of megacalyces, the following criteria must be satisfied: (1) the calyces should appear faceted; (2) the kidney must be large with multiple calyces; (3) the infundibula, pelvis, and ureter should have a normal caliber [5]; (4) there should be an absence of vesicoureteral reflux [1]; (5) the isotope renogram should demonstrate normal renal function.

IVU remains the investigation of choice for the diagnosis, because it explores the whole urinary tract and provides information about the triad calyx-ureter-renal pelvis [1].

After intravenous injection of contrast material, the time to appearance in the calyces is the same on both sides. However, because of the large volume of urine in each megacalyces there is delay in full visualization of the collecting system [5]. The calyces are uniformly enlarged. Their number is usually increased to 8-10 but may be as many as 20-25 [5,10]. The configuration of the renal pelvis is normal. The ureter is normal in caliber [5]. The amount of functioning cortical tissue is normal in uncomplicated cases. The renal cortex may appear slightly thinned, but only because it is stretched around the dilated calyces [12]. In a previous study, fluoroscopy showed absent peristalsis of the calyces and infundibula, infrequent peristalsis of the renal pelvis, and normal ureteral motility [5]. These observations can now be made by sonography or CT [9]. These findings were verified in our case.

IVU has proven to be very accurate in making the differential diagnosis from other disorders such as obstructive uropathy and intermittent hydronephrosis [3]. Common obstructive hydronephrosis, diffuse papillary necrosis, hydrocalice, staghorn calculi, and tuberculosis may cause confusion with megacalyces [5,7].

Management and Follow-up

Megacalyces should not be confused with obstructive uropathy, in order to avoid unnecessary surgical intervention [5,7]. Megacalyces is not progressive and surgical treatment is not necessary unless there are complications [8,9,12]. Treatment should be directed at controlling urinary-tract infections with appropriate antibiotics and attempting to minimize calculus formation with large fluid intake [5]. Extracorporeal shockwave lithotripsy is efficient in the treatment of lithiasis associated with megacalycosis [3].

A yearly control by IVU or CT urography (preferred) is indicated [3]. The function and appearance of the pelvicalyceal system typically remains stable in uncomplicated cases that are followed over several years [8,9,19].

CONCLUSION

Megacalyces is a rare congenital urinary anomaly characterized by nonobstructive dilated calyces, with a renal pelvis that has normal caliber. The diagnosis is confirmed by IVU or CT scan. The disease is not progressive and it does not require any surgery unless there are complications. Surveillance is recommended in noncomplicated cases.

REFERENCES

1. Mazas-Artasona L, Led Huerta A, Balmaseda Toyas C, González Pastor JF, Miranda Blanco JJ. Megacalyces or Puigvert's disease. Apropos of 2 cases. [Article in French]. *Ann Radiol (Paris)*. 1991;34(4):248-250.
2. Puigvert A. Megacalyosis: differential diagnosis from hydrocalyectasia [Article in Spanish]. *Med Clin (Barc).* 1963;41:294-302.
3. Pereira Arias JG, Gurtubay Arrieta I, Escobal Tamayo V, Ibarluzea González JG, Jorge Catalina A, Bernuy Malfaz C. Megacalycosis and lithiasis [Article in Spanish]. *Arch Esp Urol*. 1995;48(3):310-314.
4. Kimche D, Lask D. Megacalycosis. *Urology*. 1982;19(5):478-481.
5. Talner LB, Gittes RF. Megacalyces. *Clin Radiol*. 1972;23(3):355-361.
6. Vargas B, Lebowitz RL. The coexistence of congenital megacalyces and primary megaureter. *AJR Am J Roentgenol.* 1986;147(2):313-316.
7. Bekele W, Sanchez TR. Congenital megacalyces presenting as neonatal hydronephrosis. *Pediatr Radiol*. 2010;40(9):1579.
8. Kozakewich HP, Lebowitz RL. Congenital megacalyces. *Pediatr Radiol*. 1974;2(4):251-257.
9. Gittes AF. Congenital megacalices. In: Stamey TA, ed. *Monographs in Urology*. Research Triangle Park, NC: Burroughs Wellcome Co; 1984:1-20.
10. Gabutti L, Alerci M, Marone C. Spiral CT angiography for discriminating between megacalyces and intermittent hydronephrosis. *Nephrol Dial Transplant.* 1997;12(7):1487-1489.
11. Hamrick LC, Burns JR. Congenital megacalices in a black woman. *J Urol.* 1985;134(4):714-715.
12. Gittes RF, Talner LB. Congenital megacalices versus obstructive hydronephrosis. *J Urol.* 1972;108(6):833-836.
13. Marsilli E, Camerani M, Maffei I, et al. Mega-poly-calicosis. Apropos of a case with prenatal and postnatal diagnosis [Article in French]. *J Urol (Paris)*. 1991;97(6):294-297.
14. Mellins HZ. Cystic dilatations of the upper urinary tract: a radiologist's developmental model. *Radiology.* 1984;153(2):291-301.

15. Lam AH. Familial megacalyces with autosomal recessive inheritance. Report of 3 affected siblings. *Pediatr Radiol.* 1988;19(1):28-30.

16. Itatani H, Koide T, Okuyama A, Mizutani S, Sonoda T. Development of the calyceal system in the human fetus. *Invest Urol.* 1979;16(5):388-394.

17. Von Niederhäusern W, Tuchschmid D. A poorly-known association: primary congenital megaureter and megacalyx kidney [Article in French]. *Ann Urol (Paris)*. 1971;5(4):225-231.

18. O'Reilly PH. Relationship between intermittent hydronephrosis and megacalicosis. *Br J Urol*. 1989;64(2):125-129.

19. Galian P, Forest M, Aboulker P. Megacalycosis [Article in French]. *Presse Med*. 1970;78(38):1663-1667.