Case Report

Clinical progression of megacalycosis in a girl with a solitary kidney: The lesson learned

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Abbreviations & Acronyms
CRP = C-reactive protein
MC = megacalycosis
MRI = magnetic resonance imaging
SCr = creatinine
TLC = total leukocyte count
UTI = urinary tract infections

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Introduction: Megacalycosis is a rare disorder related to congenital underdevelopment of the renal papilla or structural defect of the renal calyces. Megacalycosis has a wide spectrum of clinical presentations ranging from simple variants without any significance on renal function to severe complications. Any prevention strategy is recommended yet since megacalycosis is mostly asymptomatic and usually discovered either accidentally or as result of its complications.
Case presentation: We observed megacalycosis progression in a young female with a single kidney toward progressive calyx dilatation for years, which ended in acute pyelonephritis. Conservative management, urinary drainage, and large-spectrum antibiotics were unsuccessful and nephrectomy was required.
Conclusion: This rare case and literature review add evidence to identify prognostic factors to select patients with a high risk of complications (single kidney, bilateral disease, female gender, associated genetic syndromes, vesicoureteral reflux, and contralateral renal disorder). One o more factors should activate close monitoring and prophylactic therapy if needed.

Key words: acute pyelonephritis, dialysis, megacalycosis, solitary kidney.

Keynote message

- Megacalycosis sometimes can lead to severe complications, such as urinary infections, stone formation, and end-stage renal disease.
- Acute pyelonephritis secondary to megacalycosis may be severe enough to require radical nephrectomy
- A careful follow-up may be necessary in patients with megacalycosisin order to prevent and treat promptly urinary tract infections and renal stones.

Introduction

MC is a rare congenital disorder related to abnormal development of the renal pyramids or structural defect of the renal calyces that lead to the absence of peristalsis with concomitant non-obstructive calyceal dilatation.[1–3] Its pathogenesis remains unclear. MC is mostly unilateral and asymptomatic, with a male-to-female ratio of 6:1, and Caucasians are mainly affected.[4]

MC has an uneventful course in most cases but sometimes can lead to severe complications such as urinary infections, stone formation, hematuria, progressive renal enlargement, and end-stage renal disease in some cases.[5,6] Furthermore, MC might be found in association with a wide spectrum of congenital anomalies of the kidney and urinary tract. In the presence of more than 30 calices for each kidney, this anomaly is commonly named Megapolycalycosis, but this is infrequent and did not impact clinical progression. MC is usually diagnosed or treated only when complications occur.[7] A strategy for monitoring and preventing complications has not yet been defined.

We describe a young woman with MC in a solitary kidney followed for 13 years. She developed a progressive dilatation of the renal collecting system associated with recurrent UTI, which ended in an episode of acute pyelonephritis requiring nephrectomy.

Case report

A 23-year old woman was admitted to our hospital for persistent fever and right flank pain lasting for 10 days. She was already on antibiotic therapy with Ciprofloxacin and Fluconazole for 7 days. Her medical history revealed multiple episodes of UTI treated with antibiotics in the past 2 years. She already had a MRI of the abdomen that showed a right solitary kidney with megacalicosis associated with a bicornuate uterus.

Laboratory investigations at admission revealed: hemoglobin 9.3 mg/dL, TLC 26.06*10^3/mmc, neutrophils 93%, CRP 246 mg/L, pro-calcitonin 97.2 ng/mL, random blood glucose 103 mg/dL, serum urea 96 mg/dL, SCr 2.7 mg/dL with an estimated-GFR of 20, serum Na^+ 123 mEq/L and K^+ 5.6 mEq/L. Urine samples were collected with no sign of pyuria or haematuria. Multiple urine and blood cultures collected with different timing resulted negative.

She underwent a new MRI that revealed a pyelonephritis-like aspect in the right kidney with a maximum diameter of 17.7 cm (Fig. 1a,b), severe reduction of cortical tissue thickness, and corpuscular urine in the collecting systems.

Renal MR imaging was compared with those obtained in 2009, which showed a longitudinal diameter of 11.9 cm with normal parenchymal thickness (Fig. 1c), while images obtained in 2013 revealed a renal longitudinal diameter of 12.5 cm with progressive and mild thinning of the cortical rim (Fig. 1d,e), without hydronephrosis.

Wide-spectrum antibiotic therapy was empirically started with intravenous Meropenem 500 mg four times a day and Teicoplanin 400 mg every 48 h. Abdominal non-contrast CT excluded urinary stones. A double J ureteral stent and Foley bladder catheter were positioned to drain the whole tract. Urine from the upper urinary tract was collected for culture but no pathogen was isolated. After 1 week, there was no improvement in laboratory parameters and a percutaneous nephrostomy was placed to drain the largest calyx but the patient did not improve (Fig. 2).

Fifteen days after her admission, nephrectomy was deemed necessary to control infection and performed using a right flank open approach. Cloudy urine was drained from the enlarged calyces. The surgical specimen (Fig. 3) appeared markedly enlarged with a normal pelvis and ureter. The histopathology report confirmed the diagnosis of MC associated with diffuse pyelonephritis. The renal parenchyma showed cortex and medulla markedly thinned overlying dilated calyces. An inflamed medulla and blunted papillary tip were present. The renal parenchyma showed chronic pyelonephritis with associated acute inflammation and abscess formation. There was an abrupt transition from the normal cortex, where the glomeruli were spared, to an area of scarring characterized by tubular atrophy, interstitial fibrosis, severe inflammation and globally sclerotic glomeruli.

The postoperative stay was uneventful with progressive improvement, with a SCr of 1.4 mg/dL. The patient was discharged on the 7th postoperative day. She is still on hemodialysis and waiting for renal transplantation.

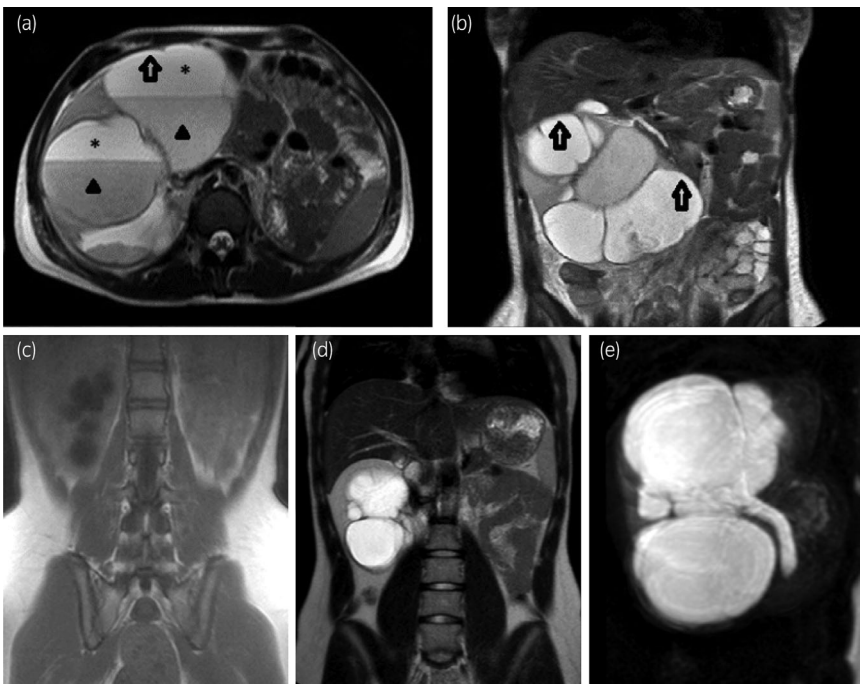

Fig. 1 (a,b) Abdominal MR at hospital admission in 2022. The solitary right kidneys occupies almost the entire right side of the abdomen dislocating the other structures. Atrophic and thin renal parenchyma (arrows), the urine in the lumen is half-filled in two parts with cell debres (arrowhead), while in the upper half is watery fluid (*). (c) MRI obtained in 2009 showing MC with a normal cortex obtained. (d) MRI obtained in 2013 showing increase of calices dilatation and cortical thinning. (e) MRI Urographyn (2013) showing normal pelvis and ureter.

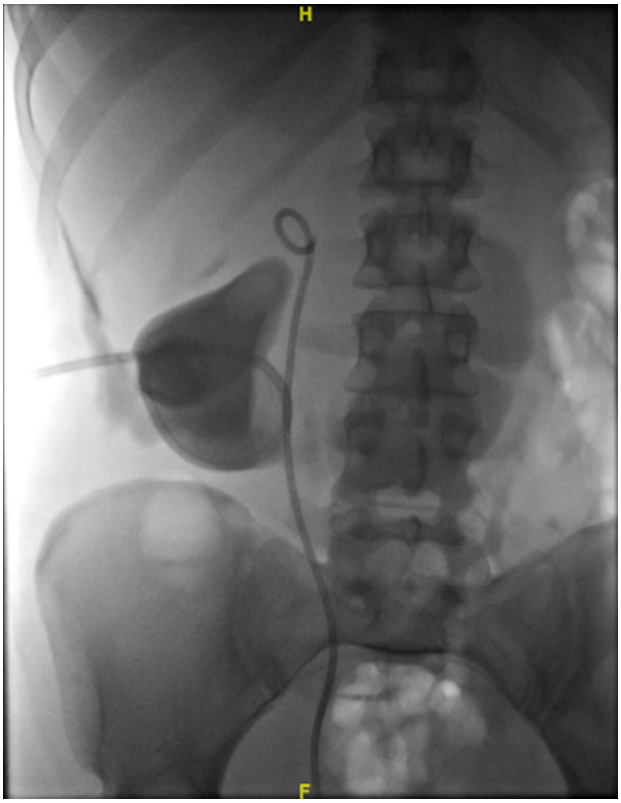

Fig. 2 Fluoroscopic image after ureteral stenting and nephrostomy placement. The lowest calyx appears dilated with a narrow infundibulum.

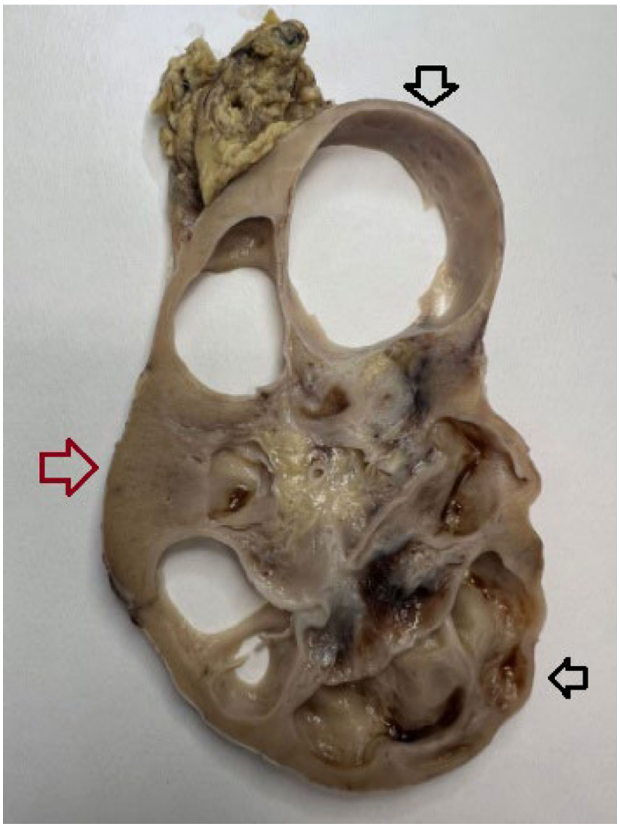

Fig. 3 Surgical Specimen of the whole right kidney: calyces appear dilated with thinned cortical tissue (black arrows), while residual parenchyma is still visible (red arrow).

Discussion

MC associated with contralateral renal agenesis is an extremely rare condition that has been described in two cases.[8,9] The first case was presented in 1981 in a 4-year-old girl with Vacterl/Vater Syndrome and multiple malformations (skeletal, gastrointestinal, and genitourinary).[8] The second case was found in 2018 in a young girl with urinary tract infection.[9] The follow-up of those patients is unknown. Conversely, we were able to follow the natural course of MC in our patient throughout 13 years and this lead us to understand what could happen in kidneys with MC over a long-term period.

Infection of the megacalyces can be commonly treated with antibiotics with a good prognosis[10] even when affecting both kidneys. Multiple episodes of UTI led probably to multi-drug resistant pathogen selection in our patient, which converted to uncontrollable pyelonephritis.

Evaluating the progressive morphologic changes of the patient's kidney through the MRI imaging, wide dilatation of some calyces progressed in time: we hypothesize that they affected renal function by calyceal compression and stretching on adjacent calyces leading to loss of urine outflow from the corresponding renal tissue. In Figure 1, atrophic and thin renal parenchyma surround exceptionally enlarged calyces. The urine in the lumen is half filled with cell debris in the low and watery liquid in the upper part. Hence, nephrectomy was the only way to eradicate her life-threatening infection.

Congenital anomalies of the kidney and urinary tract are crucial for the outcomes of complications in patients with MC.[11] Furthermore, renal function is lost only when severe complications (infection and stones) occur. Therefore, monitoring renal function is not enough. The progression of MC is unpredictable, but the prevention of complications is mandatory.

Based on the literature review and this case, we identified factors for complications: single kidney, bilateral disease, female gender, vesicoureteral reflux, megaureter, associated genetic syndromes, or other contralateral renal disorders. The presence of one o more of these risk factors should activate the adoption of close monitoring and prophylactic therapy.

In conclusion, the present case shows that calyx dilatation in MC can be progressive and lead to infective complications and renal damage. Patients diagnosed with MC should be carefully followed during their entire life to prevent and treat properly the high incidence of recurrent UTI and stones.

Author contributions

Andrea Benedetto Galosi: Conceptualization; writing – original draft; writing – review and editing. Carlotta Nedbal: Data curation; investigation. Vanessa Palantrani: Data curation; methodology. Andrea Ranghino: Project administration; visualization. Roberta Mazzucchelli: Resources. Carlo Giulioni:

Project administration; writing – review and editing. Giulio Milanese: Writing – review and editing. Daniele Castellani: Software; writing – review and editing.

Conflict of interest

None.

Approval of the research protocol by an Institutional Reviewer Board and the approval number

Not applicable.

Informed consent

The informed consent was obtained from the subject.

Registry and the Registration No. of the study/trial

Not applicable.

References

1 Puigvert A. La Megacaliosis. (Disembrioplasia De las Pir'amides De Malpighio) [Megacalycosis. Dysembroplasia of the MalphigianPyramids]. *Rev. Clin. Esp.* 1963; **91**: 69–80.

2 Anselmo G, Cunico SC. Le displasie megacalicali. *Urol. J.* 1965; **32**: 376–86.

3 Kimche D, Lask D. Megacalycosis. *Urology* 1982; **19**: 478–81.

4 Partin AW, Dmochowski RR, Kavoussi LR, Peters CA. *Urology Cambpell-Walsh-Wein* ed. Elsevier, Philadelphia,PA, 2022; 717–721, 738–739.

5 Kalaitzis C, Patris E, Deligeorgiou E *et al*. Radiological findings and the clinical importance of megacalycosis. *Res. Rep. Urol.* 2015; **7**: 153–5.

6 O'Connor CJ, Kinnear N, Browne G, Hennessey DB. Bilateral staghorn kidney stones in Megacalycosis: non operative management of complex kidney stone disease. *Urol. Case Rep.* 2022; **44**: 102146.

7 Redman JF, Neeb AD. Congenital megacalycosis: a forgotten diagnosis? *Urology* 2005; **65**: 384–5.

8 Toguri AG, Hayden K, Travis L. Link between caudal regression and VATER syndromes. *Urology* 1981; **17**: 469–72.

9 Szmigielska A, Krzemień G, Zacharzewska A, Dudek-Warchoł T, Warchoł S. Wrodzonekielichonercze u dziewczynki z jednostronnaagenezjanerki [Congenital megacalycosis in a girl with unilateral renal agenesis]. *PolMerkurLekarski* 2018; **44**: 205–7 Polish.

10 Kasap B, Kavukcu S, Soylu A, Türkmen M, Secil M. Megacalycosis: report of twocases. *Pediatr. Nephrol.* 2005; **20**: 828–30.

11 Hildreth TA, Stewart W, Cass AS. Congenital megacalyces associated with Hirschprung's disease. *Urology* 1976; **7**: 187–90.