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Clinical Case Seminar CCS 2 (1-4)

Un “uncommon” case of polycystic kidney disorder

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Abstract

An anomaly *radiologically* detected at a first line examination- if not properly contextualized - may lead to erroneous diagnostic hypotheses and subsequent therapeutic interventions. Hydronephrosis could be sometime misdiagnosed with a diffuse cystic involvement of one kidney causing parenchymal enlargement with a normal contralateral kidney. We report here a case of a 55 -year-old man with suboptimally controlled arterial hypertension and progressive increase of creatinine serum levels, who initially received a diagnostic suspicion of unilateral polycystic kidney disease. After an accurate differential diagnosis, our findings were consistent with a unilateral hydronephrosis caused by stenosis of ureteropelvic juncture due to an aberrant renal artery. Patient has undergone successfully robotic surgery, with progressive clinical and laboratory improvement.

Keywords: unilateral kidney cysts; renal failure; stenosis of ureteropelvic juncture; aberrant renal arteries

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Introduction

Renal diseases can have a wide range of clinical and biochemical correlates and imaging frequently assumes a central role in diagnostic approach. Ultrasound, due to its non-invasiveness and relative cheapness, represents the first line examination carried out when a kidney damage is suspected, leaving the other methodologies to the subsequent diagnostic steps (1).

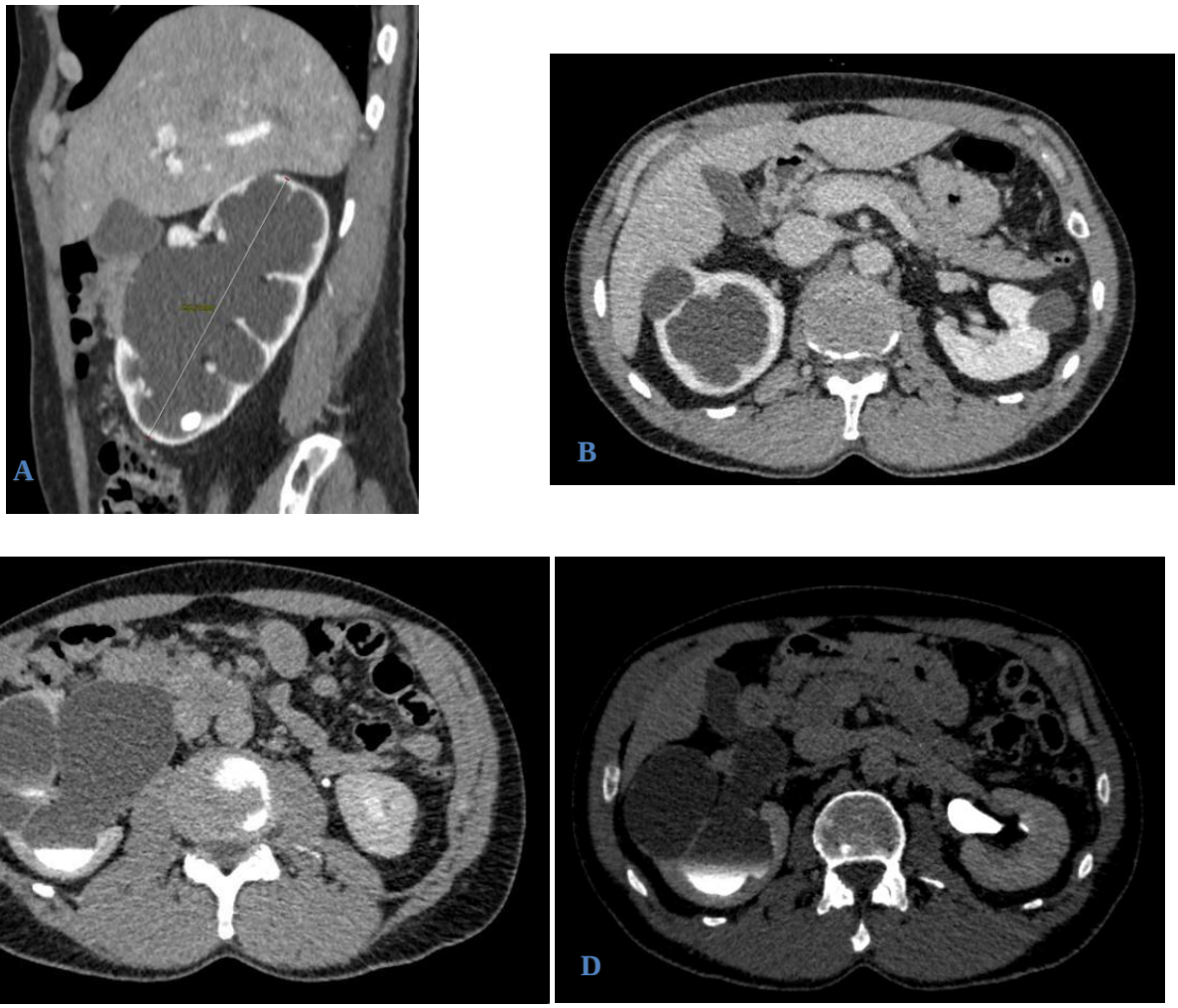
The Kidney Disease: Improving Global Outcomes (KDIGO) recently published clinical practice guideline for Autosomal Dominant Polycystic Kidney Disease (ADPKD) (2). In this paper, a diagnostic algorithm in adults with an incidental report of kidney and/or liver cysts in absence of known family history of ADPKD is proposed. Nevertheless, the clinical scenario could induce the suspicion of other concurrent pathologies, such as stone disease, solid kidney lesion or hydronephrosis. The latter is defined as dilatation and distention of the renal collector system of one or both kidneys due to obstruction of urine outflow distally to the renal pelvis (ureter, bladder, or urethra), whose clinical manifestations range from paucisymptomatic onset to abdominal/back pain, fever, weight loss and/or hematuria; causes of hydronephrosis may include kidney stones, enlarged prostate, pregnancy, urinary

tract infections, certain kinds of cancer, trauma and stenosis of ureteropelvic juncture. (3) Below, we present a case of an adult man to whom, after a thorough diagnostic approach, a unilateral hydronephrosis due to stenosis of ureteropelvic juncture secondary to an accessory renal artery was diagnosed and effectively treated with robotic surgery.

Case report

A 55-year-old man was admitted to our Internal Medicine outpatient clinic due to a gradual increase of creatinine and azotemia serum levels (latest value 1.7 mg/dl and 52 mg/dl, respectively) in the last two years and a suboptimal blood pressure control despite good adherence to oral dual therapy with once daily olmesartan 20 mg and nebivolol 5 mg. The patient had a 25 pack-years of smoking history and was affected by dyslipidemia not in pharmacological treatment and hypertension diagnosed about six years earlier. At a clinical examination there wasn't peripheral oedema, and the patient reported mild right side abdominal pain which arose a few months earlier; blood pressure value was about 145/95 mmHg. Vesicular breath sounds were slightly reduced in both mid-apical fields on chest auscultation. He had undergone abdominal ultrasound at other diagnostic center, in whose report the following was stated "… The left kidney of normal size, cysts with a maximum diameter of 2.94 cm. The right kidney increased in volume with poorly assessable size and morphology completely subverted by the presence of multiple cystic formations, the largest being 5.25 cm. A lithiasic formation of 1.23 cm in one of the cysts. Undilated excretory tract.". Referring to the diagnosis algorithm in adults with incidentally detected kidney and/or liver cysts in absence of known family history of ADPKD, proposed in the latest KDIGO 2025 ADPKD Guidelines, in presence of multiple cysts and/or kidney enlargement, reduced kidney function, liver cysts and no atypical extrarenal findings, we could have already confirmed ADPKD diagnosis in our patient, with eventually suggestion to perform genetic testing (2). It could be nosographically recognized as an "Unilateral Atypical ADPKD", according to the Mayo Imaging Classification (MIC) (2), however, symptomatological and biohumoral features - and ultrasound report - have suggested that we should move toward another diagnostic approach. Thus, contrast-enhanced abdominal computed tomography (CT) was performed, showing severe right hydronephrosis and ipsilateral stenosis of the ureteropelvic juncture, driven by abnormal contact with the inferior polar accessory renal artery (Fig 1)

Figure 1. Abdominal contrast-enhanced CT showing severe right hydronephrosis with kidney increased in volume (longitudinal diameter = 15 cm; panel A), significantly thinned parenchyma, and evident dilatation of the calyceal cavities with stenosis at the level of the ureteropelvic juncture (panel B and D), due to anomalous contact with inferior polar accessory renal artery, which runs behind the junction and bifurcates early. Three small stones (panel A) are present in the context of the calyceal cavities and significantly slowed urinary excretion is present (panel C and D).



Propaedeutic to urologic indication of right nephrostomy, a dynamic renal scintigraphy was performed, in which was detected at right kidney a slowed excretion of the tracer through the collecting system and a function of 62% and 38% for left and right kidney, respectively. Thereby, he underwent robot-assisted pyeloureteroplasty and placement of right indwelling Double J ureteral stent.

After 30 days this robotic surgery, a new dynamic renal scintigraphy detected an improved extraction of the tracer and GFR measurement at right kidney was increased to 48% (42.4 ml/min versus 29.72 ml/min). In the following days, the patient experienced occurrences of dysuria associated with flank discomfort at right side, which resulted as episodes of urinary tract infection, successfully treated with targeted antibiotics. Laboratory data carried out in the following days showed a progressive improvement of renal function indices (creatinine serum level lowered to 1.1 mg/dl) and favorable clinical outcomes: a less intensive

antihypertensive drug treatment was reached, with discontinuation of nebivolol and halving of olmesartan from 20 to 10 mg daily, obtaining a better blood pressure control (110/75 mmHg).

Conclusions

Our singular report describes a case of hydronephrosis due to abnormal contact with accessory renal artery with onset in the adult subject and few cases are reported to date in the literature (4,5).

A careful clinical approach in guiding the patient's diagnostic and instrumental process is crucial. Among differential diagnosis of the adult patient with unilateral hydronephrosis and suspected secondary hypertension, stenosis of ureteropelvic juncture should be considered. Finally, early investigations of decline of renal function can therefore allow both early diagnosis and a better therapeutic decision-making with improved patient's prognosis and quality of life.

Conflicts of Interest: There is no potential conflict of interest, and the Authors have nothing to disclose. This work was not supported by any grant.

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