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Epidemiological, Clinical and Evolutive Profile of Autosomal Dominant Polycystic Kidney Disease (ADPKD) in Togo

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Abstract

Objective: To describe the epidemiological, clinical and evolutionary profile of ADPKD in Togo. **Methods:** A retrospective descriptive transversal study over a period of 8 years (2011-2018) which focused on the analysis of patients' records diagnosed with ADPKD. The diagnosis of ADPKD was retained on the basis of the ultrasound criteria of PEI. **Results:** During the study period, 27 patients had polycystic kidney disease with a prevalence of 0.87%. The average age was 51.6 ± 16.4 years. There were 10 men (37%) and 17 women (63%), a sex ratio (M/F) of 0.58. The concept of family cystic kidney disease was found in 6 (22.2%) patients. The clinical presentations were dominated by arterial high blood pressure, abdominal pain and abdominal mass respectively in 77%, 63% and 63% of cases. Five patients (18.5%) had a glomerular filtration rate (GFR) greater than 90 ml/min, 17 (62.9%) had a GFR < 60 ml/min. All patients (100%) had multiple renal cysts, 16 patients (59.3%) had dedifferentiated kidneys. Six patients (22.2%) had liver cysts, one patient (3.7%) had lithiasis. Genetic was not achieved because of the poor technical platform and the high cost of these tests. **Conclusion:** ADPKD is common in our department. It appears to be associated with a high rate of chronic renal failure.

Keywords

Polycystic Kidney Disease, CKD, Togo

1. Introduction

ADPKD is a chronic multi-systemic disease of hereditary origin, characterized

by the development of cysts in both kidneys as well as by variable extrarenal organ manifestations [1]. It affects one birth in 800 or 4 to 6 million people around the world [2]. This is an autosomal dominant inheritance disorder. Therefore, the father or mother of the patient with ADPKD also carries the disease. Autosomal dominant inheritance implies that members of the same family are affected in each generation, with the risk of transmitting the disease from the affected parent to the child at 50%, regardless of gender. ADPKD is caused by mutations in the PKD1 gene (located on chromosome 16p13.3) or PKD2 (chromosome 4q21) [1].

In Togo, ADPKD is relatively common in hospitals. Our study is the first in Togo. At the time of the development of various therapies more or less effective on the ADPKD [3], it seemed important TOUS to describe the epidemiological, clinical and evolutive profile of the ADPKD patients of our hospital population.

2. Patients and Method

Our study was conducted in the nephrology and hemodialysis department of the Sylvanus Olympio University Hospital of Lomé in Togo. This department, which is the only public center for the treatment of kidney diseases in the country, also carries out care and research activities. This is a retrospective cross-sectional descriptive study from January 1st, 2010 to December 2017, which is an 8-year period that focused on the analysis of patients with ADPKD diagnosed cases. The diagnosis of ADPKD was retained on the basis of the ultrasound criteria of PEI [4]. The kidneys were considered dedifferentiated if the renal cortex appeared hypoechogenic relative to the medulla. Included, were all patients aged 15 years and more who are being followed-up at the Sylvanus Olympio University Hospital in Lomé for ADPKD. Patients who did not meet the PEI ultrasound criteria and patients under 15 years of age were not included. The concept of family cystic kidney disease has also been investigated in the interrogation of patients diagnosed with ADPKD. The clearance of serum creatinine has been estimated using the Modification Diet of Renal Disease (MDRD) formula [5]. Patient data were collected using charts with socio-demographic, clinical, biological and ultrasound parameters. This data was analyzed and processed using Sphinx Plus software (Lexica), version 5.1.0.3.

Definition of PEI criteria: The presence of three or more (unilateral or bilateral) renal cysts is sufficient for establishing the diagnosis in individuals aged 15 to 39 years, two or more cysts in each kidney is sufficient for individuals aged 40 to 59 years, and four or more cysts in each kidney is required for individuals ≥60 years. Conversely, fewer than two renal cysts in at-risk individuals aged ≥40 years are sufficient to exclude the disease.

3. Results

During the study period (2010-2017), 3064 patients were received in the Department of Nephrology and hemodialysis of the Sylvanus Olympio University Hos-

pital of Lomé. Twenty-seven patients had polycystic kidney disease with a prevalence of 0.87%. Among the 27 patients, there were 10 men (37%) and 17 women (63%) a sex ratio of 0.58. Table 1 showed the sociodemographic characteristicaspect. The concept of family cystickidney disease was found in 6 (22.2%) patients. The average age was 51.6 years ± 16.4 years with extremes of 25 and 91 years. One patient (3.7%) was carrying Human Immunodeficiency Virus (HIV), two (7.4%) were sickle cell. The genetic study was not performed.

3.1. Clinical Manifestation

The clinical presentations were dominated by high blood pressure, abdominal pain and abdominal mass respectively in 77%, 63% and 63% of cases. Table 2 shows the distribution of patients according to the clinical manifestations of polycystic. Four patients (14.8%) had electrocardiographic left ventricular hypertrophy. Three patients (11.1%) had died.

3.2. Biological Parameters

The mean hemoglobin level was 10.6 g/dl ± 2.6 g/dl with extremes of 5.7 and 14.2 g/dl.

The mean serum calcium was 88.4 mg/L ± 11.9 mg/L with extremes of 60 and 124 mg/l. The mean proteinuria of 24 h was 501 mg/day ± 458 mg/day with extremes of 84 and 2 g/day. Five patients (18.5%) had a glomerular filtration rate (GFR) greater than 90 ml/min, two patients (7.4%) had a GFR < 15 ml/min but not yet in hemodialysis and five patients (18.5%) were in iterative hemodialysis at two sessions per week. Table 3 shows the distribution of patients according to creatinine clearance.

3.3. Echographic Parameters

The diagnosis of polycystic kidney disease is based on clinical arguments, the notion of family cystic kidney disease and especially on the ultrasound arguments of the disease. All patients (100%) had multiple kidney cysts, 16 patients (59.3%) had dedifferentiated kidneys. Table 4, Table 5 shows the distribution of

Table 1. Sociodemographic and clinical characteristics of the patients.

	Effective	%
Sex		
Male	10	37%
Female	17	63
Profession		
Actives	22	81.4
Pensionners	4	14.8
Student	1	3.7
Comorbidities		
HIV	1	3.7
Sickle cell	2	7.4

Table 2. Division of patients according to clinical manifestations.

	Effective	%
Abdominal pain	17	63.0
HBP	21	77.8
Macroscopic hematuria	6	22.2
Urinary infection	7	25.9
Abdominal mass	17	63.0

HBP: High Blood Pressure.

Table 3. Patient Division Based on Creatinine Clearance (MDRD).

	Effective	%
GFR ≥ 90	5	18.5
90 > GFR ≥ 60	5	18.5
60 > GFR ≥ 30	5	18.5
30 > GFR ≥ 15	5	18.5
GFR < 15 (waiting for dialysis)	2	7.4
GFR < 15 (dialysis)	5	18.5
Total	27	100

GFR: Glomerular Filtration Rate.

Table 4. Division of patients according to ultrasound parameters of polycystic kidney disease.

	Effective	%
Multiple renal cyst	27	100
Dedifferentiated kidneys	16	59.3
Liver cyst	6	22.2
Pancreatic cyst	0	0.0
Splenic cyst	0	0.0
Renal lithiasis	1	3.7

Table 5. Division of patients according to the circumstances of discovery.

	Effective	%
Adventitious	4	14.8
CKD	10	37.0
Lumbar-abdominal pain	8	29.6
Other: hematuria, urinary system infections	5	18.5
Total	27	100

patients according to the ultrasound parameters of polycystic kidney disease. Six patients (22.2%) had liver cysts, one patient (3.7%) had renal lithiasis. None of

our patients had splenic or pancreatic cysts.

3.4. Evolution

In our series, 3 patients (11.1%) had died two of whom had infectious complications and one a stroke. The mean follow-up time for deceased patients was 2.6 years. The two patients who died of infectious cause were 44 years old and 45 years old. The one who died of a stroke was 50 years old.

4. Discussion

ADPKD is a hereditary disease that, because of its autosomal dominant inheritance, affects about half of the family members. In our series the notion of family cystic kidney disease was found in 6 (22.2%) of our patients. BOURQUIA *et al.* [6] reported that the family survey allowed early diagnosis of ADPKD. The diagnosis of mutation remains resolutely the responsibility of genetics. In our current practice this analysis is not carried out because of the lack of adapted material and also because of the high cost of these tests.

The average age of our patients was 51.6 ± 16.4 years. RAQUI *et al.* [7] reported an average age of 48.8 ± 16.6 years; N'GUESSAN in 2015 in Côte d'Ivoire [8] had observed in his patients, an average age of 47.5 ± 13.8 years. We observed a sex ratio of 0.8. N'GUESSAN *et al.* [8] in their series reported a sex ratio of 1. Similarly in Senegal, out of 55 subjects with PKD, 57% were men [9]. It appears that there is no real predominance of sex.

ADPKD is a serious condition because of its multiple complications, which most often represent the reason for consultation and the circumstances of discovery. In this study the reasons for consultation were dominated by kidney failure (37%), and lower back pain (29.6%), while in Senegal [9] the most common circumstances of discovery were lower backpain (52.2%) and high blood pressure (17.4%). The most common clinical manifestation in our series was high blood pressure (77% of cases). High blood pressure is the most common complication of ADPKD. It was found in 56% of cases in the N'GUESSAN series [8] and in 36% of cases in Senegal [9]. This high blood pressure appears early, already when the GFR is still greater than 75 ml/min, compared to the general population [10]. This observed difference can be explained by the fact that high blood pressure is more common in kidney failure, especially when the patient has a family history of high blood pressure. High blood pressure during ADPKD is associated with increased renal volume [1]. Serra *et al.* [1] were able to show that in patients with ADPKD, aged 15 to 49 years, with a GFR greater than 60 ml/min/1.73 m^2, who, takes ACE inhibitors, were within a target blood pressure range of 95/60 to 110/75 mm·Hg, the increase in renal volume was slower [1].

CKD during ADPKD is very common since about 17 patients (62.9%) in our series had a CKD and among them two (7.4%) had an ESRD but not yet on hemodialysis and five patients (18.5%) an ESRD in iterative hemodialysis. The proportion of polycystic hemodialysis patients in our departement was 6.1%,

whereas it is generally around 10% [11]. This work complements polycystic kidney disease in Africa. Although there is no consensus in the diagnosis, monitoring and treatment of this pathology [11], some new therapies have emerged. In our present work we have not addressed the therapeutic aspect of polycystic kidney disease.

In Benin, AGBOTON *et al.* [12] in their series noted that 75% of patients had stage III chronic kidney disease (CKD). EL HADJ FARY KA in Senegal noted that 51% of patients were CKD, the majority being in stage I, followed by dialysis [13]. These very high rates of CKD can be explained by the late use of patients for care because of their ignorance of the disease but also because of very poor socio-economic conditions. Indeed, the guaranteed minimum inter-professional salary being 58€ (SMIG = 38,000 FCFA), taking care of oneself is a real problem for patients in Togo. It should also be noted that the absence of universal health insurance contributes enormously to the lack of health care.

Six patients (22.2%) in our series had macroscopic hematuria. Hematuria is very common during ADPKD and occurs at least once in life in half of patients. Although it is often a source of great concern, it is usually benign. More rarely, it is due to an infection of the urinary system. As far as urinary infections are concerned, the danger is that, from there, the infection can spread to the kidney itself. There is a risk that a cystic kidney infection treated too late or insufficiently renders a nephrectomy essential, thus greatly reducing renal function [1].

ADPKD is characterized by the slow and progressive appearance of cysts mainly in the kidneys. It can be associated with the development of cysts in the liver, spleen and pancreas. Extrarenal manifestations in our study were mainly dominated by liver cysts (22.2%). GOMEZ *et al.* observed in their study that the most frequent extra-renal manifestations were liver cysts (62%) and pancreatic cysts (9%) [14]. This observed difference can be explained by the quality of the ultrasound scan used.

In this study, 3 patients (11, 1%) had died including 2 infectious complications and 1 stroke. In fact the prevalence of intracranial aneurysms is about 10% (compared to 2% - 3% in the general population) [1]. The incidence of cerebral bleeding associated with aneurysms, however, is low. A general screening is not recommended by most experts and is reserved for special situations, for example to pilots, before a planned transplant, or to families with frequent aneurysms.

5. Conclusion

ADPKD is common in our department. It appears to be associated with a high rate of chronic renal failure. Systematic screening of the affected family members should be encouraged for early referral of patients to nephrologists to provide comprehensive and long-term care for patients and relatives.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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