PERICARDIAL EFFUSION ASSOCIATED WITH POLYCYSTIC KIDNEY DISEASE – CASE REPORT

DERRAME PERICÁRDICO ASSOCIADO A DOENÇA RENAL POLICÍSTICA – RELATO DE CASO

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ABSTRACT

Introduction: Autosomal dominant polycystic kidney disease (ADPKD) is a comorbidity with a progressive evolution, with the development of renal cysts and consequent loss of renal function. It is considered the fourth disease that leads to kidney failure in the world. Extra-renal symptoms are common, such as the presence of cysts in other organs, valve abnormalities, vascular changes, aneurysms, pericardial effusion, among others. Objective: The objective of the study is to associate ADPKD as a cause of pericardial effusion through a report of a case report. This is a descriptive observational study of a resident of the city of Valença-RJ, Brazil, male, treated at the Medical Clinic Ward of the Hospital Escola de Valença -RJ. Brazil Results: Patient with ADPKD presenting pericardial effusion to be clarified, with the final causal relationship polycystic kidney disease itself. Conclusion: After the literature review and the case report, it was evident that pericardial effusion is a common clinical finding in patients with Polycystic Kidney Disease, but little correlated, mainly because it is asymptomatic in the majority of cases.

KEYWORDS: Polycystic kidney disease, systemic arterial hypertension, pericardial effusion.

RESUMO

Introdução: A Doença renal policística autossômica dominante (DRPAD) é uma comorbidade com evolução de caráter progressivo, com desenvolvimento de cistos renais e consequente perda da função renal. É considerada a quarta doença que mais leva a falência renal no mundo. Sintomas extra-renais são comuns, como a presença de cistos em outros órgãos, anormalidades valvares, alterações vasculares, aneurismas, derrame pericárdico, entre outros Objetivo: O objetivo do estudo é associar a DRPAD a como causa do derrame pericárdico por meio de um relato de um relato de caso. Este é um estudo observacional descritivo de um morador da cidade de Valença-RJ, Brasil, sexo masculino, atendido na Enfermaria de Clínica Médica do Hospital Escola de Valença –RJ.Brasil Resultados: Paciente com DRPAD apresentando derrame pericárdico a esclarecer, tendo como relação causal final a própria Doença renal policística. Conclusão: Após a revisão bibliográfica e o relato de caso, evidenciou-se que o Derrame pericárdico é achado clínico comum em pacientes com Doença renal policística, porém pouco correlacionado, principalmente por ser assintomático na maioria dos casos.

PALAVRAS-CHAVE: Doença renal policística, hipertensão arterial sistêmica, derrame pericárdico.

1. INTRODUCTION

Autosomal dominant polycystic kidney disease (ADPKD) is a comorbidity with a progressive course, with the development of kidney cysts and consequent loss of kidney function. It is considered the fourth disease that leads to kidney failure in the world[1].

Approximately 50% of individuals with this comorbidity progress to the final stage of chronic renal failure around the age of 60, a development that is due to the size of the renal cysts and destruction of the parenchyma[1].

The clinical picture of most patients with polycystic kidney disease is asymptomatic. A small proportion will develop early symptoms. Most patients are diagnosed due to a family history or development of refractory arterial hypertension[2].

The increase in kidney size and the activation of the renin-angiotensin-aldosterone system are mainly responsible for the development of systemic arterial hypertension in cases of polycystic kidney disease. As a result, most of these patients have left ventricular hypertrophy as a result of increased systolic blood pressure. The main cause of death in patients with polycystic kidney disease is cardiovascular disease, with arterial hypertension being the main determinant for the unfavorable prognosis [2] There is no laboratory test or exam to diagnose ADPKD, but the clinic associated with imaging tests has been a great ally at the time of diagnosis[1].

The disease is diagnosed later in males than in

females, resulting in a worse prognosis for the disease in men. This is a contributing factor to the greater number of patients on dialysis due to polycystic kidney disease being men[3].

Extra-renal symptoms are common, such as the presence of cysts in other organs, valve abnormalities, vascular changes, aneurysms, pericardial effusion, among others[4].

Pericardial effusion is a common clinical finding, mainly related to cardiovascular diseases. It may be related to both intra and extracardiac changes. In most cases it is asymptomatic and is characterized as a find. The causes are diverse, such as inflammatory, infectious, immunological, physical, hormonal and rheumatological. Non-cardiac causes such as neoplasia, trauma, kidney injury or even pregnancy are also described[5].

It is known that there is a close association between the occurrence of pericardial effusion and polycystic kidney disease. Its presence frequently occurs in polycystic kidney patients as an extrarenal sign, where most cases are asymptomatic. Pericardial effusion occurs exponentially more frequently in the population with polycystic kidney disease compared to the general population. Women are more affected than men[6].

This causal relationship can be explained. Two genetic defects are linked to polycystic kidney disease. The PKD1 gene defect is the most common, seen in 85% of cases, while PKD2 is involved in 10 to 15% of the remainder. The pathophysiology of pericardial effusion in this disease is not yet fully known, however it is believed to be more related to the PKD1 gene, which generates the abnormal production of components of the extracellular matrix of the connective tissue, increasing compliance and decreasing pericardial return[7].

Another factor to be considered in polycystic kidney disease is the increased production of plasma renin, which consequently causes harm and makes blood pressure control difficult, with greater rapidity in relation to the decline in kidney function[8].

The aim of the study is to associate ADPKD as a cause of pericardial effusion through a case report.

This is a descriptive observational study of a male resident of the city of Valença-RJ,Brazil, treated at the Medical Clinic Ward of the Hospital Escola de Valença -RJ. Brazil carried out a brief review of the topic for the theoretical basis of the article in databases such as PubMed, Scielo and UpToDate. Data collection was carried out through analysis of the patient's medical records and medical evolution. In accordance with the ethical precepts for research with humans.

2. CASE REPORT

Patient, 46 years old, brown, rural worker, sought medical attention with reports of edema of the lower limbs up to the knee region bilaterally associated with burning pain of a mechanical nature and swelling of the right calf that had started 15 days ago. Previously hypertensive for 10 years, with polycystic kidney disease using losartan 100mg/day and hydrochlorothiazide 25mg/day. Family history being that of a mother who died of chronic kidney disease on dialysis. He denied smoking. Social alcoholism. Upon admission physical examination, the patient presented with anasarca, hypertensive (BP 150x120 mmHg), saturating 90% in room air and tachypnea, however, without respiratory effort. Globose abdomen showing bilaterally palpable polycystic kidneys. Lower limbs were swollen with asymmetry, with the right lower limb being more notable, hyperemia and swelling of the calf in addition to 3cm more circumference compared to the left lower limb. Upon admission to the ward, a chest x-ray was requested showing an enlarged cardiac area. Electrocardiogram with sinus rhythm, amplitude alternation between QRS complexes and ST rectification on the lateral wall. Admission laboratory tests were requested showing kidney injury. Due to the increase in the cardiac area on a chest X-ray and a history of polycystic kidney disease associated with kidney injury, it was decided to request a chest CT scan and USG (ultrasonography) of the kidneys and urinary tract. USG with venous Doppler of the lower limbs was also requested to rule out peripheral thrombotic disease.

USG of the lower limbs with venous Doppler/arterial Doppler: no evidence of deep vein thrombosis. Total abdominal tomography: kidneys with increased dimensions and lobulated contours due to multiple hypodense cortical formations, some with fine peripheral calcifications and others with slightly hyperdense content, the set of findings is compatible with adult polycystic kidney disease; liver cysts; Chest tomography: massive pericardial effusion, atelectatic streaks/subsegmental atelectasis are noted in the lung parenchyma adjacent to the pericardial effusion. USG of kidneys and urinary tract shows evidence of polycystic kidneys. Transthoracic Echocardiogram: Pericardial effusion, mild mitral valve insufficiency, left ventricle with significant degree of concentric hypertrophy. Laboratory tests: Hemoglobin: 16.2, leukocytes: 7,600, platelets: 154,000, urea: 68, creatitine: 2.6, sodium: 143.

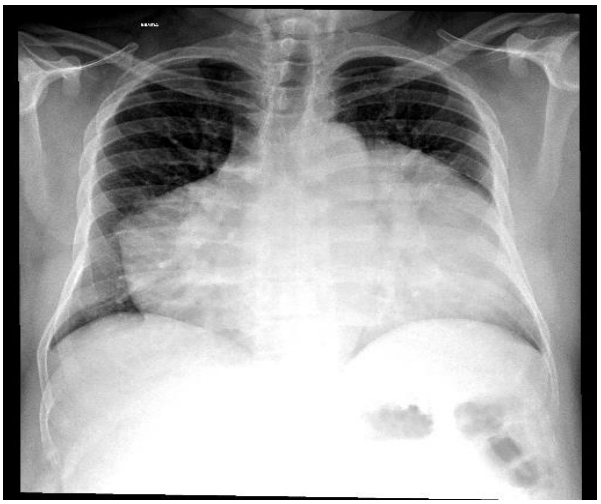

Figure 1. PA chest x-ray. Presence of increased mediastinal area.

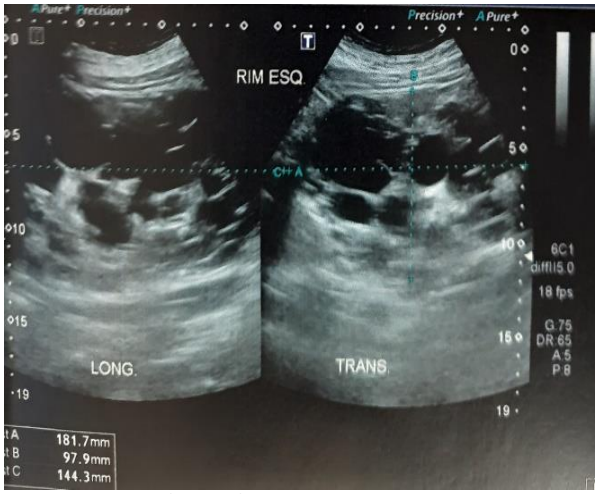

Figure 2. USG of the left kidney showing round anechoic areas suggestive of multiple renal cysts.

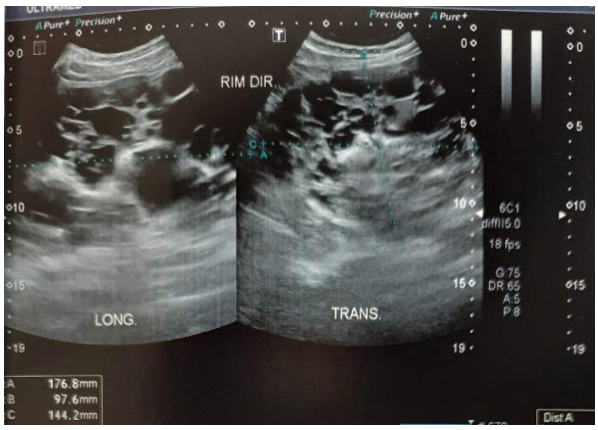

Figure 3. USG of the right kidney. showing round anechoic areas suggestive of multiple renal cysts.

Five days after admission, USG-guided diagnostic and relief pericardiocentesis was performed. Drained 1700ml of citrine yellow liquid. An analysis of the pericardial fluid was performed, showing no signs of infection through a negative culture, cellularity 300 cells/mm3 with 80% lymphocytes, 20% neutrophils; gram test without growth, protein 4.4g/dl, glucose 114g/dl, PH 7, lactic dehydrogenase 331 u/l. Uric acid was evaluated in both serum and pericardial fluid under a microscope, where the presence of uric acid crystals was not visualized. Adenosine deminase dosage test and oncotic cytology of pericardial fluid were negative, ruling out neoplastic and tuberculosis processes. Thyroid evaluation tests were carried out and the causal thyroid factor was excluded. The patient did not present any rheumatological clinical signs or symptoms. After draining the pericardial effusion, the patient began experiencing symptomatic hypotension (BP: 90x70 mmHg) where it was necessary to discontinue antihypertensive medication as the patient was using amlodipine 10mg/day. Colchicine and associated prednisone 20mg/day were maintained empirically as the renal function did not tolerate NSAIDs for the treatment of pericarditis, even without laboratory evidence of an infectious condition. Five days after pericardiocentesis, the patient continued to experience edema and return of difficult-to-control hypertension, reaching a maximum hypertensive peak of 200x126 mmHg. Loop diuretic was started due to anasarca and lower limb edema. After 7 days of hospitalization, the patient lost 4.1 kg with partial improvement in lower limb edema. In the following days, diuretic therapy with triple blockade (furosemide, chlorthalidone and spironolactone) was optimized. The patient remained with refractory hypertension despite the use of optimized antihypertensive medications, namely: Spironolactone 25 mg/day, chlorthalidone 25 mg/day, clonidine 0.900 mg/day, hydralazine 300 mg/day, amlodipine 10 mg/day, carvedilol 12.50 mg/day, isosorbide 120 mg/day, furosemide 40 mg/day, methyldopa 1g/day. On the twenty-first day, the patient is discharged from the hospital with refractory hypertension (BP 170x100 at the time of the visit), with guidance for early outpatient reassessment, having ruled out the main causes of pericardial effusion as being of infectious, thyroid, rheumatological, or uremic origin, neoplastic, medicinal, gastro-intestinal. Referred to a nephrology and cardiology outpatient clinic with optimized prescription, maintaining the use of antihypertensive drugs from the last hospital prescription, diuretic therapy and colchicine 1 mg/day for 3 months associated with prednisone for 3 weeks, being advised to wean as follows: prednisone 20 mg 1 and ½ cp in the morning for 7 days, increasing to a dose of 1cp/day for 7 days, decreasing to 1/2cp a day for 7 days, ending with prednisone 5mg/day for 4 days, ceasing use. The patient continued to be monitored at the cardiology and nephrology outpatient clinic, remaining asymptomatic and with blood pressure values below 140x90mmHg measured at the time of the consultation and bedside USG showing the presence of moderately intense pericardial effusion.

3. DISCUSSION

Although there was no diagnostic confirmation by genetic material in laboratory analysis, it is noted that the patient in the reported case falls into the age group most affected by individuals with polycystic kidney disease associated with a maternal family history of chronic kidney disease. dialysis, in addition to confirmation by imaging tests. There is a correlation between the epidemiological profile of ADPKD and the profile of the patient in question. (two).

Pericardial effusion has been a common finding in patients with chronic polycystic kidney disease. Often a finding that is little noticed, or is wrongly justified by another etiology, forgetting the genetic-cellular and structural change caused by Polycystic Kidney Disease in the various tissues and structures of the human body[9].

Furthermore, it is noted in literature reports that there are major extrarenal repercussions of ADPKD, exemplified in the patient in question. During complementary exams, cystic formations in the liver, large-volume pericardial effusion, mild degree of mitral insufficiency, and left ventricular hypertrophy were

revealed. These findings further corroborate the impacts of polycystic kidney disease beyond the renal system[1].

The pathophysiology of ADPKD is increased production of plasma renin by juxtaglomerular cells. Thus, there is an increase in systemic blood pressure. These mechanisms elucidate the refractory arterial hypertension of the patient in the case report, since despite being medicated with several antihypertensives from different classes, he maintained high blood pressure. This explains why the patient has refractory hypertension. (two)

4. CONCLUSION

After the literature review and the case report, it became clear that pericardial effusion is a common clinical finding in patients with polycystic kidney disease, although it is poorly correlated, mainly because it is asymptomatic in most cases. In short, evidence of pericardial effusion does not exclude other pathologies as its cause and its relationship can be made by excluding even more common causes of this condition. Another point that can be addressed is refractory hypertension, already well explained and correlated in patients with polycystic kidney disease, unlike pericardial effusion, which despite its correlation already being well demonstrated, still requires further studies to prove it from a pathophysiological point of view.

5. REFERENCES

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