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Giant polycystic kidney disease: Treatment with open bilateral nephrectomy - A case report with successful outcomes

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Abstract

We present the case of a 53-year-old male with a family history of Polycystic Kidney Disease (PKD) and a 20-year diagnosis of PKD who refused surgical treatment and opted for alternative medicine. After a month on hemodialysis, he presented to the emergency department with chest pain, dyspnea, somnolence, desaturation, hypotension, abdominal pain, and gross hematuria. Hemodynamic stabilization was performed, followed by an urgent bilateral nephrectomy due to cardiovascular repercussions. A subcostal incision approach was used, revealing a left kidney weighing 7 kg and a right kidney weighing 9 kg with multiple cysts. This case highlights the importance of timely intervention.

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Keywords: Giant polycystic kidney disease; Open bilateral nephrectomy; Chronic kidney disease; End-stage chronic kidney disease.

Introduction

PKD is a progressive systemic disorder characterized by multiple bilateral cysts in the renal parenchyma [1]. It is the most common hereditary kidney disease with a global prevalence ranging from 1:400 to 1:1,000 and affecting approximately 12 million people worldwide [2]. The progressive development of cysts may replace renal parenchyma and aggravate renal function, leading to the development of End-Stage Chronic Kidney Disease (ESCKD) in the late phase of the disease that requires renal replacement therapy or hemodialysis [3,4]. The affected kidneys frequently grow to a larger size, causing compressive symptoms such as abdominal distension or pain, back pain, nausea, and early satiety. Complications include recurrent gross hematuria from hemorrhagic cyst, recurrent urinary tract infections from infected cyst, nephrolithiasis, hypertension, and predisposition to malignancy. About 20% of patients eventually have their kidneys removed [5].

Case presentation

A 53-year-old male with a family history of 2 siblings with Polycystic Kidney Disease (PKD) and a 20-year history of PKD who refused surgical treatment and opted for alternative medicine. He had been on hemodialysis for one month because of ESCKD. He presented with chest pain, dyspnea, somnolence, desaturation, hypotension, abdominal pain, and gross hematuria. Physical examination revealed abdominal distension and bilateral flank tenderness (Figure 1). Laboratory tests showed elevated serum creatinine levels and a drop in hemoglobin levels needed blood transfusions. Hemodynamic stabilization was performed and CT scans revealed bilateral renal enlargement

measuring 42 x 35 cm right and 40 x 28 cm left with numerous large cysts, some are of hemorrhagic content (Figure 2). After hemodynamic stabilization we decide to realize an urgent open bilateral nephrectomy due to cardiovascular repercussions; using a subcostal incision, revealing a left kidney weighing 7 kg and a right kidney 9 kg with multiple cysts, with abscesses in both upper renal poles of approximately 40 cc and an abscessed hematoma on the posterior surface of the right kidney of approximately 100 cc and there was no evidence of viable renal tissue (Figures 3-5), having a surgical time of 1 hour 30 minutes and bleeding of 1000 cc requiring transfusion of blood components. The patient was transferred to the intensive care unit for postoperative care. Histopathological examination confirmed the diagnosis of polycystic kidney disease.

The patient had a good recovery and was discharged on postoperative day 14, continuing hemodialysis 3 times a week for regular follow-up and has been included in our hospital's kidney transplant program.

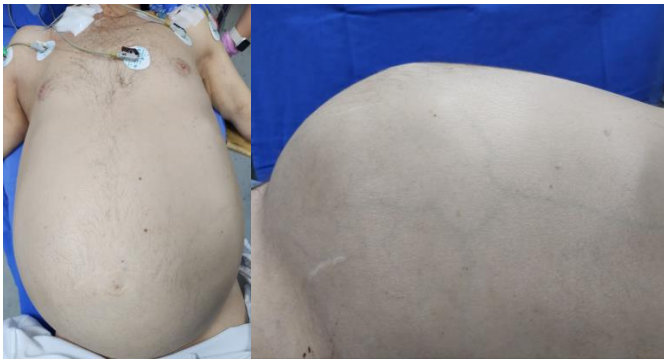

Figure 1: Anteroposterior and lateral views of the abdomen demonstrating abdominal distention.

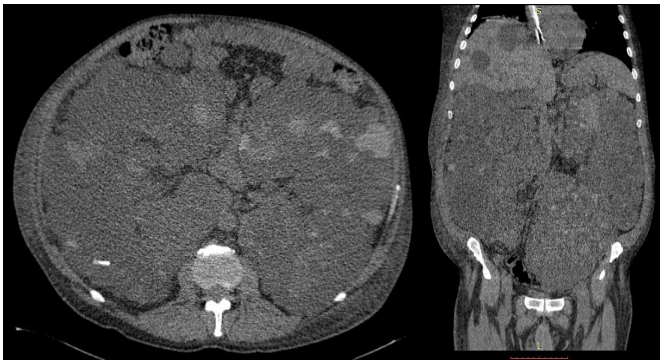

Figure 2: Abdominopelvic CT scan of multicystic kidneys with the almost complete destruction of the parenchyma.

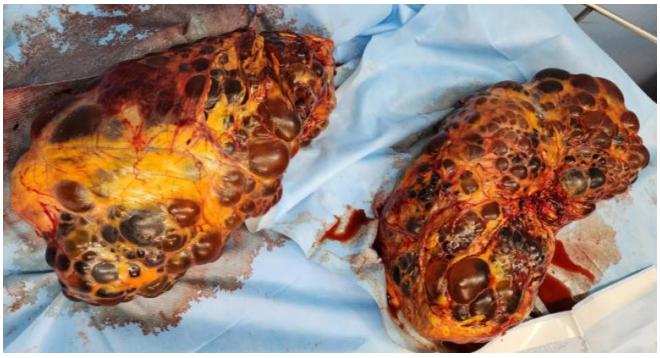

Figure 3: Macroscopic aspect of left kidney weighing 7 kg and right kidney 9 kg with multiple hemorrhagic cysts.

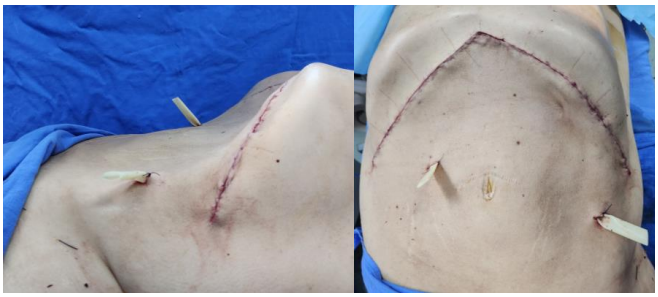

Figure 4: Anteroposterior and lateral views of the abdomen in the immediate postoperative.

Figure 5: A photograph of the highly skilled surgical team whose collaborative efforts made this operation possible.

Discussion

PKD is a leading cause of ESCKD, affecting up to 12 millions of patients. This progressive and unfortunately incurable condition can lead to significant morbidity and kidney failure. As the population undergoes more imaging for other problems, the number of diagnosed asymptomatic cases is also rising [6]. PKD was first described in 1841 by Pierre Rayer and officially named "polycystic kidney disease" in 1888 by Felix Lejars. It is the most common inherited form of chronic kidney disease worldwide. Around 12 million people globally suffer from PKD, making it more prevalent than sickle cell disease, Down syndrome, cystic fibrosis, hemophilia, and Huntington disease combined. In about 90% of new diagnoses, patients have a family history of autosomal dominant PKD7.

PKD is a multisystem disorder presenting with many kidney and extra-kidney manifestations (Table 1) [6,7].

Table 1: Summary of kidney and extra-kidney manifestations of ADPKD.

Kidney manifestations:
1. Functional changes: Concentration defect (nocturia, polyuria and frequency).
2. Hypertension.
3. Pain syndrome (caused by infection, bleeding, cyst rupture, stones or compression).
4. Uremic manifestations with declining GFR.
Extra-kidney manifestations:
1. Polycystic liver disease.
2. Intracranial aneurysms.
3. Cardiac involvement and valvular heart diseases.
4. Other cystic involvement of different organs; seminal vesicles, pancreas, etc.

Diagnosis primarily relies on imaging studies of the kidneys. Ultrasound Scan (USS) is the initial modality used for screening due to its safety, effectiveness, and affordability. Additionally, CT and MRI scans can be employed. Simple measurements of kidney length, width, and depth obtained from CT or MRI can provide a reasonably accurate estimate of total kidney volume. This estimation is calculated using the formula for an ellipsoid ($\pi/6 \times length \times width \times depth$) [8-10].

Conservative management with medical therapy is crucial. This includes controlling hypertension, maintaining hydration, reducing dietary sodium intake, and managing dyslipidemia to slow the progression of renal disease [10]. Kidney transplantation is the preferred type of renal replacement therapy for patients with PKD. Survival rates for both patients and transplanted kidneys are excellent, offering better long-term outcomes compared to hemodialysis, even when considering PKD recipients versus non-PKD recipients [11].

For pain relief, several minimally invasive procedures can be performed to decompress cysts. These procedures, cyst aspiration and sclerosis, are typically guided by ultrasound or CT and are routinely performed by interventional radiologists. A more invasive approach includes laparoscopic or surgical cyst fenestration through lumbotomy or flank incision. Kidney denervation is another option for pain management [12].

Bilateral nephrectomy is associated with a high rate of postoperative complications (38%) and a risk of mortality (3%). The surgery can be performed either open or laparoscopically, depending on the size of the kidneys and the surgeon's experience. The indications for bilateral nephrectomy include: massive enlargement of the kidneys causing abdominal symptoms (pain and early satiety), need for space for a future kidney transplant, Infected cysts, relapsing gross hematuria and some times suspected renal cancer [13,14].

Conclusion

This case report demonstrates the severe complications that can arise from PKD. While the patient initially opted for alternative medicine, the progressive nature of PKD ultimately led to ESKD requiring hemodialysis. The patient then presented with life-threatening symptoms due to massive kidney enlargement and infection. Urgent bilateral nephrectomy, although a high-risk surgery, proved successful in removing the diseased kidneys and alleviating the patient's symptoms. This case highlights the importance of early diagnosis and management of PKD to prevent severe complications and emphasizes kidney transplantation as the preferred treatment option for patients with PKD-related kidney failure.

Declarations

Data access statement: Data supporting this study are included in the article and/or supporting materials.

Funding statement: No funding was received for this study.

Research: This work was conducted under the supervision of Dr. Josué Omar Hernández Martínez as a case study under no formal organization.

Ethical compliance: All procedures performed in this study involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Conflict of interest: The authors declare that they have no affiliations with or involvement in any organization or entity with any financial interests in the subject matter or materials discussed in this manuscript.

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