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Case Report

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First case of simultaneous liver-kidney transplant in a patient with primary hyperoxaluria in the state of Santa Catarina - Brazil

Resume

Primary hyperoxaluria (PH) is a rare disorder, resulting from an excessive production of calcium oxalate mainly by the liver, which can be deposited in different organs. Patients affected by PH can progress to recurrent lithiasis, nephrocalcinosis and end-stage renal disease, requiring a kidney transplant for the recovery of organ function, associated with liver transplantation, to correct the source of this metabolic defect.[1]

The case of MJB, female, 27 years old, was reported in this paper, which is the first recorded case of simultaneous transplantation of this nature in the transplant service of Hospital Santa Isabel - Blumenau and consequently the first in the state of Santa Catarina, Brazil. She discovered the disorder through genetic sequencing at the age of 24.[2]

The patient had already been on renal replacement therapy, hemodialysis, for 4 years. Due to the severe complications of the disease, simultaneous kidney and liver transplantation was indicated. Considering the evolution of the patient up to the present moment, the transplant in this case proved to be a very effective therapeutic alternative, and in agreement with the literature.[1,3]

Keywords: liver-kidney transplant, primary hyperoxaluria, renal tubule, enzyme deficiency

Volume 11 Issue 2 - 2023

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Received: June 17, 2023 | **Published:** June 28, 2023

Introduction

Primary hyperoxaluria is a rare autosomal recessive metabolic disease, with the disorder of oxalate overproduction being the main cause of the disease. Consequently, the appearance of lithiasis and nephrocalcinosis is common, since oxalate, in the form of calcium salt, is too insoluble and excreted almost exclusively by the kidneys, tending to precipitate in the renal tubules and explain the complications.

Primary hyperoxaluria Type I, which is more frequent, has as its main characteristic the high urinary excretion rate of gluconate, caused by a low level of peroxisomal liver enzyme alanine-glyoxylate aminotransferase (AGT). Therefore, the patient's treatment is a very relevant factor for the decision on the conducts taken in cases of Primary Hyperoxaluria, in view of this, the estimated glomerular filtration rate (eGFR) is the main component evaluated, if it is less than 30 ml/min/ 1.73m2 the transplant is considered. Since the disease is closely associated with the presence of oxalate crystals in the kidney and is caused by an enzyme deficiency, kidney transplantation should preferably be associated with liver transplantation to correct the enzyme deficiency.[3]

The simultaneous transplant is efficient in preventing the recurrence of the kidney disease and can reverse the systemic accumulation of oxalate, which does not happen when the single kidney transplant is performed, which even with intensive dialysis in the pre- and post-transplant and in the use of pyridoxine, the reappearance rate and graft survival after 3 years is 15 to 25%.[4] Therefore, the present study aims to report the first case of SIMULTANEOUS liver and kidney transplantation due to primary hyperoxaluria at Hospital Santa Isabel de Blumenau - SC. Since this is one of the main liver transplant services in the country, being the 4th Brazilian hospital with the highest number of liver transplants performed in the year 2022.[5]

Case report

Female patient, 27 years old, born in Urussanga-SC, admitted to the Transplant Service of the city of Blumenau on May 2, 2022. Diagnosis of chronic kidney disease under genetic sequencing confirmed in November 2018. No kidney biopsy. Non-reactive IgG and IgM serologies for toxoplasmosis, non-reactive IgG for cytomegalovirus, non-reactive IgM and VDRL, non-reactive anti-HIV, non-reactive anti-HCV, non-reactive Anti-HBs and non-reactive HBsAg.

On renal replacement therapy since May 2017, on hemodialysis for 4 years. On the day of admission, he had creatinine levels of 11.48 mg/dL and urea of 162 mg/dL. 16 U/L TGO and 7 U/L TGP. Direct bilirubin 0.09 mg/d and indirect 0.23 mg/dL. History of acute thrombosis of arteriovenous fistulas in upper limbs. On peritoneal dialysis since 2021, 9 hours daily, 9 cycles, showing yellow and green bags. In daily use of Carvedilol 25mg 12/12h and Entresto 12/12h.

Abdominal computed tomography with findings of medullary nephrocalcinosis, retroperitoneal and intraperitoneal lymphadenopathy. Also, on 5/2/22, a SIMULTANEOUS kidney and liver transplant was performed. Through the Gibson incision, the left kidney was implanted on the left side of the patient, with immediate diuresis and uneventful surgery. Through the median "J" incision, an orthotopic liver transplant was performed using the Piggyback technique, preserving the inferior vena cava. The repercussion was excellent, fast and homogeneous, with bile production in the late phase, without intercurrences. Post-transplant laboratories presented creatinine levels of 1 mg/dL. 25 U/L TGO and 19 U/L TGP. Direct bilirubin 0.18 mg/d and indirect 0.25 mg/dL. The patient was discharged on the 30th postoperative day.

Discussion

The lack of case reports of simultaneous liver and kidney transplantation due to primary hyperoxaluria highlights the rarity

and complexity of this clinical condition. The dearth of work in the area underscores the need to share clinical experiences and treatment outcomes to better understand the efficacy and safety of this complex intervention. This case report seeks to contribute to the growing literature by providing additional information on the successful clinical application of simultaneous liver and kidney transplantation in a patient with advanced primary hyperoxaluria, highlighting the importance of this therapeutic option in clinically challenging situations.[6]

It is important to emphasize that primary hyperoxaluria (PH) is a complex and extremely rare condition, difficult to diagnose. Considering that the patient may present numerous clinical symptoms until the disease is suspected. There are numerous associated factors that contribute to the difficult approach of this disease, such as the specific type of disease, severity of symptoms, age of onset and response to treatment.

PH affects several organs and aspects of metabolism, but mainly changes the functionality of the kidneys and for this reason many patients suffer from kidney failure and need a kidney transplant. However, it is not uncommon for graft rejection to occur after kidney transplantation, as glyoxylate aminotransferase (AGT) deficiency is caused by the liver.

Therefore, simultaneous transplantation of liver and kidney is the most indicated treatment for the disease. Therefore, we present a case of a patient with primary hyperoxaluria and chronic renal failure who underwent simultaneous liver and kidney transplantation on May 2, 2022. Even with the high number of transplants performed at Hospital Santa Isabel since 1980, this was the first to be registered with this indication, being a significant milestone for the history of the hospital and the state and also in the improvement of techniques and knowledge of professionals working in the same field.[7]

In the case of primary hyperoxaluria type 1 (PH1), which represents the most common form of the disease, if the appropriate and individualized treatment is not obtained, PH can progress rapidly and result in renal manifestations at an early age.

In the past, before more effective treatments were developed, many of the patients with this condition developed end-stage kidney disease during childhood or adolescence. Which in turn resulted in a significantly reduced survival rate in those individuals who did not undergo medical intervention.

The prevalence of primary hyperoxaluria in Brazil and worldwide is difficult to estimate. It is estimated that it affects about 200 people in Brazil, including adults and children,[2] data from France in 1995 reported a prevalence of 1.05/million, with an incidence rate of 0.12/million/year, being considered a very serious rare disease.[8,9] Emphasizing the importance of more knowledge about this complex disorder.

Conclusion

Primary hyperoxaluria, being a rare disorder and difficult to estimate, is still little discussed and experienced in clinical practice. It is extremely important to be aware of its repercussions on different organs, which can lead to serious complications. Thus, simultaneous transplantation has been indicated and has proven to be a very effective alternative in preventing the recurrence of kidney disease and reversing the systemic accumulation of oxalate in the kidneys, ensuring a significant survival for patients.

In summary, although primary hyperoxaluria is a chronic and complex genetic condition, advances in early diagnosis and therapeutic options have improved the lives of patients who suffer daily from these conditions. The collaboration of physicians, researchers and patients is fundamental to guarantee the improvement in the understanding of the disease, developing new therapeutic strategies and aiming to offer a better prognosis. For its therapy, it was found that liver transplantation provides the functional enzyme alanine-glyoxylate aminotransferase, making it an effective treatment option for patients with severe HP1. As well as kidney transplantation, which is essential for the treatment of chronic kidney disease present in this group.

Associating adequate treatment, a healthy lifestyle and specialized medical follow-up, the survival rate of patients with primary hyperoxaluria has improved year after year. However, it should be noted that each case is unique, and what is essential for proper management is the individualization of treatment, as well as agility in diagnosis and therapeutic planning, optimizing results. It is important to note that in the current scenario, with increasingly specialized technologies, prognoses tend to evolve more appropriately and quickly over time, as new therapies and approaches are tried and additional research is carried out to better understand the disease.

Therefore, it is essential that patients with primary hyperoxaluria patients receive specialized primary care from the onset of their symptoms and are in contact with health professionals who are up-to-date on the most recent medical advances currently in the treatment of this condition.

Acknowledgments

None.

Conflicts of interest

Authors declare that there is no conflict of interest exists.

Funding

None.

References

1. Rios John Fredy Nieto, Zuluaga M, Higuita LMS, et al. Primary hyperoxaluria diagnosed after kidney transplantation: report of two cases and review of the literature. *Brazilian Journal of Nephrology*. 2017;39(4):462–466.
2. Hospital santa isabel rede santa catarina. Specialty transplants. 2023.
3. De almeida, José Bruno. Primary oxalosis diagnosed after kidney transplantation case report. *Brazilian Journal of Transplantation*. 2007;10(3):779–781.
4. Borges Karen Caires. Double liver and kidney transplantation: case report and literature review. 2013.
5. Brazilian transplant registry, abto. 2023.
6. Transplantation of the liver. 3rd edn. 2014.
7. Bari K, Sharma P. Optimizing the selection of patients for simultaneous liver kidney transplant. *Clin Liver Dis*. 2021 Feb;25(1):89–102.
8. Lalaguna Mallada P, García Bodega O, Castejón Ponce E, et al. Primary hyperoxaluria. *An pediatr barc*. 2004;60(4):376–377.
9. Lorenzo V, Torres A, Salido E. Primary hyperoxaluria. *Nefrologia*. 2014;34(3):398–412.