

A Polycystic Kidney Patient as Marginal Donor: Case Report

Marjinal Donör Olarak Bir Polikistik Böbrek Hastası: Olgu Sunumu

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ABSTRACT

Polycystic kidney disease is a rare autosomal dominant hereditary disorder which can progress to end stage kidney failure. In this disease, the cysts in distinct sizes and distinct number are seen in kidneys and in liver. Nowadays, there is a significant difference in the number of donors and the patients waiting for transplantation. Because of that, using marginal donors may be necessary. In this article, we aimed to discuss the usability of a 55 years old brain death polycystic kidney disease patient's organs for transplantation or not.

Key Words: Polycystic kidney disease, brain death, marginal donor

ÖZET

Polikistik böbrek hastalığı, son dönem böbrek yetmezliğine kadar ilerleyebilen otozomal dominant kalıtımı olan nadir bir hastalıktır. Bu hastalık, böbreklerde ve karaciğerde değişik büyüklüklerde multipl kistlerle seyredir. Günümüzde organ nakli için bekleyen hastalar ile organ donörleri arasında sayısal olarak önemli bir fark vardır. Bu durumda marjinal donör kullanılması gerekebilir. Bu yazıda, 55 yaşında beyin ölümü gerçekleşen polikistik böbrek hastalığı olan donörün organlarının transplantasyon için kullanılabilirliğini tartışmayı amaçladık.

Anahtar Kelimeler: Polikistik böbrek hastalığı, beyin ölümü, marjinal donör

Introduction

Autosomal dominant polycystic kidney disease (PKD), is an hereditary disease having multiple cysts in the kidneys and causing end stage kidney failure (ESRF) with deterioration in renal functions in years. Progression to ESRF is rare in the patients under 40 years, but dialysis necessitating ESRF frequency rises around 60 years old. Male gender, rise in renal sizes and hypertension occurring in early ages increases ESRF frequency. In the patients who progresses to ESRF, hemodialysis or transplantation is treatment options of priority. In these patients; frequency of hepatic cysts, intracerebral aneurysms and cardiac diseases increases (1,2). The best treatment of ESRF patients is renal transplantation. To improve the success rate; the surgeons want to present the best organ to the recipients. But in the literature, there are cases about the transplantation of PKD patients' kidneys (3,4).

In our article, we aimed to discuss a patient becoming donor for hepatic and corneal tissue but not for kidneys.

Case Report

In a 55 years old male patient was accepted with weakness complaint in his right arm and right leg, we have learnt he had PKD and hypertension in his personal history. In his family story he had 3 brothers with PKD diagnosis. The patient has been using antihypertensive therapy for two years, and he had coronary bypass operation 1 month ago for 2 coronary arteries. In the patient's examination in the emergency room; consciousness confused, heart rate 80 beats/minute, Blood pressure (BP) 130/80 mmHg and pulse oximetry value SpO₂ was 95%. Muscle strength was 2/5 in his right side. In his diffusion magnetic resonance imaging (MRI) (Figure 1a), diffusion restriction was seen 90x60 mm in left fronto-temporo-occipital region and 42x26 mm in

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the right occipital region (Figure 1a). Mannitol 20% 1g/kg was given then 0,25 mg/kg q6h was used. In computerized tomographical (CT) imaging of his abdomen, the kidneys were doubled in size bilaterally and their dimensions were 20x12 cm in right kidney and 23x1 cm in left kidney. And there were multiple cysts the biggest one was 5 cm diameter in the kidneys bilaterally (Figure 2a). In the liver, there were multiple cysts the biggest one is 15 cm diameter (Figure 2b). In the transthoracic echocardiography, ventricular ejection fraction was measured as 40-45%. After 8 hours of patient's admittance to the intensive care unit, the condition of the patient worsened, then the patient was intubated and mechanically ventilated. The patient was reevaluated; wide haematoma and edema were seen in the infarction area (Figure 1b). Noradrenaline infusion was started to the patient because his values were HR

(Heart rate) 115 beats/minute, invasive BP (Blood pressure) 70/37 mmHg. Then his vital findings got HR 84 beats/minute, BP 105/60 mmHg, SpO₂ 96%, body temperature 36 °C, his pupils were fixed in dilatated position, and the patient was unresponsive to the painful stimuli. The patient was evaluated in terms of brain death. The patient's mannitol therapy was stopped. After 12 hours brain death tests were applied. Also, apnea test was positive, and the patient was accepted as brain death. The meeting with family was positive for using the organs for transplantation. The patient's urination >100 ml/hour, blood urea values got lowered (Table 1). The patient's cornea and hepatic tissues were used transplantation, but his kidneys weren't accepted for transplantation even though its normal function (Table 1,2) because of their sizes and the patient's age and having hypertension.

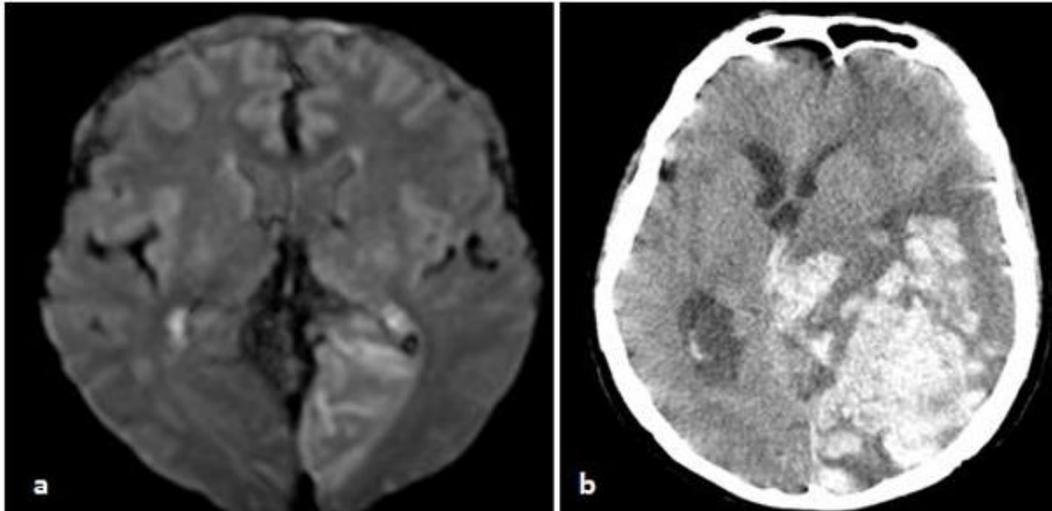


Fig. 1a). Magnetic resonance image demonstrating the cerebral ischemic infarct **b).** Computed tomographical image demonstrating the cerebral hemorrhagic infarct.

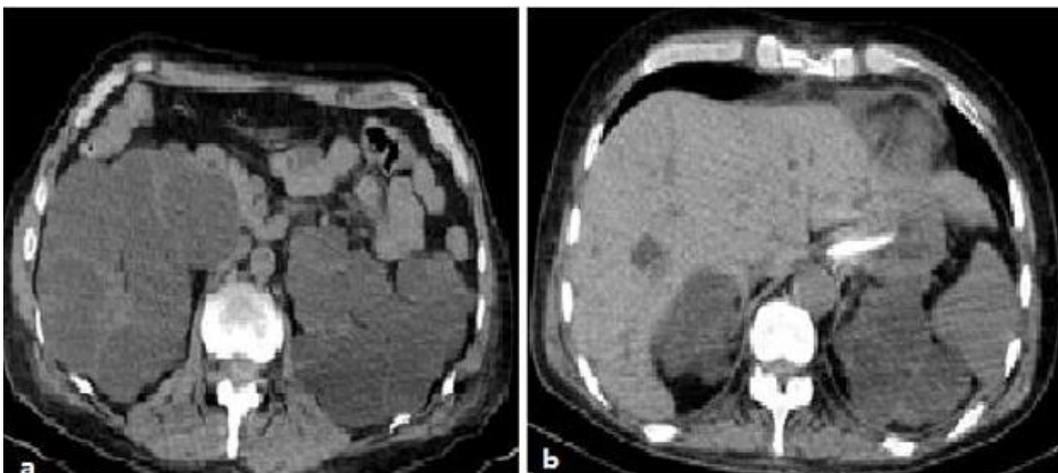


Fig. 2a). Computed tomographical image demonstrating the polycystic kidneys **b).** Computed tomographical image demonstrating the polycystic liver.

Table 1. Biochemical parameters

	4 years ago	1 month ago	3 days ago	Brain death
Creatinine	1.27	1.27	1.93	1.86
BUN (mg/dL)	42.9	39	61	71

Table 2. Urine analysis

Protein	Trace
pH	6.5
Density	1.015
Colour	Light yellow

Discussion

According to the datas of August 2017; 24807 people are waiting for transplantation in our country 21600 of them are waiting for renal transplantation. In the last 10 years, 30021 organ transplantation were performed. 100% of unblock transplantation was obtained from cadavers, 67% of standard transplantation and 97% of split transplantation was performed from live donors. Despite 12170 brain death diagnosis in the last ten years, 69% of probable male donors and 73% of probable female donors' relatives haven't agreed to donation. In today's condition, there is a serious gap between demand and supply. It has an importance to use marginal donors (cadavers, well controlled HT, diabetes, low-medium proteinuria and donors older than 60 years old) in our low leveled organ donation country (5-7).

It is a reality PKD is in the etiological causes of 10-15% of ESRF. But the beginning of PKD symptoms and the progression to renal failure length is about 10 years. When this fact is considered, though the progression probability to ESRF of 50 years old PKD patients 23%, but the progression probability to ESRF of 70 years old PKD patient is about 48% (4,5).

According to Turkish Nephrology Society, 15000 patients are having ESRF diagnosis per year and more than 25000 patients have dialysis therapy. According to 2004 recordings, 5,3% of hemodialysis because of ESRF is related to PKD. The quality of life of the patients' having ESRF with any cause gets worse and there are lots of complications awaiting them (2,3).

In a study performed in a London center, 24, 46 and 55 years old 3 PKD patients' kidneys were used for kidney transplantation between 2000-2004. In this study it is said that 8 PKD patients' kidneys have been used for transplantation until now. They found that average age of 8 PKD

patients was 23 about in their study they found it was 45 years old. It was seen that pyelonephritis occurred in one of the patients, the other two patients' renal function in follow up of 2 and 5 years were good. The main feared states of the PKD donor patients are fast deterioration of renal functions, hemorrhage in the kidney, infection and stone formation in the kidney. Renal cell carcinoma of the kidney of PKD patients is similar to the general population and it is not necessary to fear of renal cell carcinoma formation (5,8).

The issue of whether the cysts of transplanted kidneys are getting bigger or not is not clear. It is guessed that 1% of nephrons in the polycystic kidney becomes cystics because of that the kidney transplanted should have enough functional capacity. A scoring system is used for the kidneys which will be transplanted, microscopic evaluation in four ways is made. If glomerulosclerosis, tubular atrophy, interstitial fibrosis and vascular disease (arteriolar hyalinosis) are present, transplantation failure probability is high. If glomerulosclerosis >20% in biopsy, it has relation to poor graft. In a study made for PKD donors; donor's age being 50 years old, the kidney being <15 cm, performing kidney biopsy before transplantation, being donor's creatinine values normal, being recipients' life expectancy lower than 10 years and giving information about PKD to the recipients were emphasized (7-9).

The organ donation ratio is very low in developing countries like ours (Turkey), because of that it is very important to use PKD patients' organs as marginal donors if the recipient's life expectancy is short, and the donor's kidney sizes and functions are normal.

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