Kidney transplantation from a living donor to a mentally disabled recipient with bilateral angiomyolipomas—A case report

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ABSTRACT

INTRODUCTION: Many centers do not perform transplantation in mentally disabled people. Our patient with progressive psychomotor developmental delay had bilateral angiomyolipomas.

PRESENTATION OF THE CASE: Three years ago she underwent a right nephrectomy for massive spontaneous hemorrhage. The left kidney had a large, well-vascularized angiomyolipoma ready at any moment to bleed spontaneously was functioning normally. Two renal transplantation centers in Croatia refused to transplant from the patient’s donor mother. The transplantation team had concerns whether to transplant a kidney to a person unable to care for herself, about who would take complete care of the patient, including regular immunosuppressive therapy, and whether it was ethically justified to explant a functioning kidney, although affected by angiomyolipomas, from a patient who required no renal replacement therapy at the time.

CONCLUSION: We presented a successful kidney transplant in a mentally disabled person, clinical and ethical justifications for such a procedure, and a four-year post-transplant evaluation. Furthermore, in our opinion, renal transplantation in the mentally challenged needs to be referred to in literature exclusively as a relative contraindication instead of an absolute one, as has been practiced to date. This would facilitate transplantation teams deciding on kidney transplantation in mentally incapacitated individuals.

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1. Introduction

Kidney transplantation is now the globally accepted best treatment of patients with end-stage renal disease [1−3]. Although all patients are entitled to this kind of treatment and in spite of the provisions of law, treating mentally disabled persons by transplantation encounters resistance in many renal transplantation centers, partly because severe psychiatric illness and significant mental retardation are reported as absolute contraindications for kidney transplantation [4,5]. The question arises as to what treatment should be used in patients with mental disablement, epilepsy and tumorous renal malformations [6].

Tuberous sclerosis is a rare autosomal dominant hereditary disease occurring due to a chromosome 9q34 or, in some families, chromosome 16 defect that causes mental retardation [7]. Multiple nodular tumors are composed of abnormal neuronal and glial, occasionally calcified, cells found in the cortex and in other parts of the brain. Progressive mental retardation and epileptic seizures develop in early childhood and are associated with a range of skin changes that resemble white spots and angiofibroma-adenoma sebaceous in a butterfly pattern on the cheeks, chin and forehead as the clinical hallmark of the disease [8]. With the progression of the disease angiomyolipomas, tumorous malformations, may occur in the kidneys, liver, adrenal gland and pancreas. Mentally disabled patients with epilepsy and such tumors rarely live longer than 30 years. Transplantation in a mentally disabled person is not a surgical problem; it is a problem because postoperative care is risky and complex [9].

2. Presentation of the case

The work has been reported in line with the SCARE criteria [10]. A young girl, born in 1984, was admitted for work-up and opinion on the feasibility of kidney transplantation in 2006 [11,12]. The patient was diagnosed with developmental delay, including mental retardation and epilepsy in her early childhood. At the age of 4.5 months she was first hospitalized for seizures, diagnosed with West’s syndrome, and recommended antiepileptic therapy. A diagnosis of tuberous sclerosis was made in 1992. MRI of the brain showed visible native as well as post contract
MRI of the brain showed typical changes characteristics for tuber-
sous sclerosis with multiple hematomas cortically, subependimally, 
peri- and intra-ventricularly. Because of the arrest of psychomotor 
development, the patient was under the care of rehabilitation insti-
tutions. In July 2003 she was admitted to the urology department 
of another hospital for angiomyolipomas of both kidneys. Several 
days later an urgent nephrectomy of the right kidney with massive 
blood replacement was performed for massive spontaneous bleed-
ing from the angiomyolipoma. Hemorrhages from the left renal 
angiomyolipoma, which continued to grow, occurred on several 
occasions afterwards and were treated by blood transfusion. The 
left kidney was therefore functioning with angiolipomatous lesions 
and the patient was at a high risk of spontaneous hemorrhage and 
bleeding to death.

The patient’s mother volunteered to donate a kidney [13]. The 
standard pre-transplant workup protocol revealed no con-
traindications for organ donation. Tissue typing showed negative 
lymphocytotoxic antibodies, HLA phenotypes A2, A25, B35, B51, 
DR4, DR11 and DQ3. Both the recipient and the donor were of 
0 Rh positive blood type [14,15], and MSCT angiography showed 
one artery per kidney. Regarding family genetics TSC1 and TSC2 
genesis mutations was not found. Two renal transplantation centers 
in the Republic of Croatia refused to perform the transplantation, 
although the patient’s mother wished to donate a kidney.

The transplantation team had the following concerns: first, a 
dilemma whether to transplant a kidney to a person unable to care 
for herself; second, who would continue taking complete care of 
the patient, including administering regular immunosuppressive ther-
apy; and, third, whether to explant a kidney that was functioning 
although affected by angiomyolipomas, from a patient who at the 
time required no renal replacement therapy, was ethically justified. 
Because of the relatively high risk of severe blood loss and death, 
as well as the patient mother’s big motivation to donate the organ, 
we decided to perform a transplantation and carried out the pre-
transplant work-up. After undergoing a work-up at the Depart-
ment of Nephrology, the patient was transferred to the Urology Division 
of the Department of Surgery on January 9, 2006.

The findings of the preoperative workup performed on admis-
sion were within normal ranges: leukocyte count 5.09 × 10⁹/L, 
red blood cell count 9.93 × 10¹²/L, hemoglobin 114 g/L, hemat-
ocrit 0.37/L, potassium 4.1 mmol/L, sodium 140 mmol/L, chloride 
104 mmol/L, blood glucose 5.7 mmol/L, urea 4.0 mmol/L, creatinine 
111 mmol/L, and C-reactive protein 2.0 mg/L. Abdominal and pelvic 
CT scans demonstrated a large angiomyolipoma filling the entire 
left retroperitoneal space up to the entrance of the small pelvis, 
invading the renal parenchyma, compressing the pyleoclesal sys-
tem and enclosing the left ureter. Contrast-enhanced scan did not 
reveal extravasations, hemorrhages nor hematomas.

One transplantation team used a transabdominal approach to 
perform a left nephrectomy in the recipient via median laparotomy. 
Pathohistology findings showed adipose tissue, smooth muscle 
component, thick walled blood vessels and abnormal tumor ves-
sels. Also, we did not find any elements of malignant alteration. The 
other team simultaneously explanted the donor’s kidney using a 
lumbar approach. The explanted donor kidney was then implanted 
into the recipient’s left iliac fossa. Cold ischemia time was 58 min 
and warm ischemia time was 35 min. The arterial anastomosis was 
sutured end-to-side to the external iliac artery using 6-0 prolene 
and the vein was sutured end-to-side to the external iliac vein by 5-0 
prolene [16,17]. The ureter was sutured to the urinary bladder using 
the Lich–Gregoir technique with a double JJ stent insertion [18].

After the surgery, the patient was transferred to the intensive 
care unit (ICU) and stayed intubated and mechanically ventilated 
until the following morning. Laboratory test results were satis-
factory, except for the C-reactive protein, which was 85 mg/L. 
Immunosuppression was continued (cyclosporine, mycophenolate 
mofetil, corticosteroids) with antibiotic and antifungal protec-
tion, H2 blockers, and Pneumocystis carinii prophylaxis [19,20]. 
The patient received immunosuppressive therapy, anticoagulant 
therapy, antileptics, antipsychotics, antibiotics and other support 
therapy if needed. Further, the patient was circulatory stable and 
with profuse diuresis. Volume replacement with colloids and crys-
talloids was carried out using analgesic and sedative medications. 
The patient was extubated on her second day at the ICU. The 
kidney ultrasound was satisfactory, with the correction of ane-
mia; CMV prevention was carried out and antiepileptic drugs 
were given per os. The second day after the transplantation lab-
atory findings were as follows: blood glucose 7.8 mmol/l, urea 
5.6 mmol/L, creatinine 131 mmol/L, potassium 4.4 mmol/L, sodium 
146 mmol/L, chloride 118 mmol/L, calcium 1.26 mmol/L, and mag-
nesium 0.46 mmol/L. The patient gradually recovered at the ICU and 
was transferred to the nephrology division after six days.

The patient visited another transplantation center in her town of 
residence for follow-up examinations and our center for annual 
check-ups. Four years after the transplantation renal function was 
normal and the patient’s parents are taking care to adhere to 
immunosuppressive and other therapies. The ultrasound of the 
kidney graft four years after transplantation showed the kidney 
size was 127 mm, the width of parenchyma being approximately 
16 mm, and that of the cortex 7–9 mm. The cortico-medullar margin 
was clearly visible, parenchyma was of normally ecographic struc-
ture and there were no signs of concretions. The canal system was 
not significantly dilated, with the pyelon transverse diameter being 
about 18 mm. The ultrasound also showed no perirenal collections.

3. Discussion

The successful transplantation and a functioning kidney are the 
best evidence that the decision to perform the surgery was correct. 
The first two concerns about whether to transplant a kidney to a 
mentally disabled person unable to care for herself and about fur-
ther care of the patient were successfully resolved. Although many 
centers do not accept kidney transplantation in mentally disabled 
patients, it needs to be pointed out that everyone has an equal right 
to treatment. If a mentally disabled person requires dialysis, taking 
care of regular immunosuppressive therapy after transplantation 
is much more feasible to guardians than taking an uncooperative 
patient to hemodialysis two or three times a week.

The macroscopy and pathohistology of the removed kidney 
answered our third concern as to whether a still functioning 
angio-lyomipomatous kidney should be removed and what the risk 
of spontaneous bleeding was. The tumor size was 25 × 16 × 10 cm 
(Fig. 1). The entire parenchyma was invaded with yellowish-rosy 
tumor with areas of hemorrhage, and the canal system was 
passable, but and compressed (Figs. 2 and 3). The tumor was his-
tologically composed of mature fatty tissue, blood vessels with 
thickened walls and radially distributed spindle-shaped smooth
4. Conclusion

In conclusion, by performing this procedure and by following-up the patient with an adequately functioning kidney transplant on a regular and long-term basis supported by family care for the patient’s compliance with immunosuppressive and other therapy, we have demonstrated that renal transplantation in mentally insufficient patients should become a routine practice rather than constitute the controversy among transplantation professionals. Furthermore, in our opinion, renal transplantation in the mentally challenged needs to be referred to in the literature exclusively as a relative contraindication instead of an absolute one as has been practiced to date. This would facilitate transplantation teams deciding on kidney transplantation in mentally incapacitated individuals.

Conflict of interest

The authors of this manuscript have no conflict of interest to disclose as described by International Journal of Surgery Case Reports. The Results presented in this paper have not been published previously in whole or part.

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Ethical approval

The manuscript is not a clinical trial, but the surgical procedure performed on the patient. In accordance with that we didn’t need any Ethical Committee approval.

Consent

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Author contribution

Željko Vidas, surgery procedures, the conception and design of the study, drafting the article and revising it critically for important intellectual content, final approval of the version to be submitted.

Franjo Jureneč, surgery procedures, the conception and design of the study, drafting the article and revising it critically for important intellectual content, final approval of the version to be submitted.

Eva Lovrič, histopathology analyses, the conception and design of the study, drafting the article and revising it critically for important intellectual content, final approval of the version to be submitted.

Marko Samardžija, the conception and design of the study, drafting the article and revising it critically for important intellectual content, final approval of the version to be submitted.

Registration of research studies

The manuscript is not a clinical trial, but the surgical procedure performed on the patient. In accordance with that we didn’t need any Ethical Committee approval.

Guarantor

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