Renal Transplantation in a Patient with Polycystic Horseshoe Kidney - Case Report

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Abstract

Polycystic horseshoe kidney is a very rare occurrence. A 47-year-old woman patient was admitted to hospital complaining of increased belly volume. Familial history was positive for horseshoe kidney and polycystic kidney. Computerized tomography showed enlarged kidneys with multiple cysts, fused by their lower poles and multiple liver cysts. She remained on dialysis for four years, being submitted to kidney transplantation with a related live donor, presenting normal renal function after nineteen months of follow up.

INTRODUCTION

Horseshoe kidney results of an anomaly fusion during embryogenesis, while the polycystic kidney is an autosomal dominant hereditary disorder. It is a very rare occurrence found both anomalies in the same patient. The incidence ranges of 1 in 134,000 to 1 in 8,000,000 live births [1].

CASE PRESENTATION

A 47-year-old woman was admitted to hospital complaining of increased belly volume. At physical examination, palpation of large lobulated mass occupying the entire abdomen. An abdominal ultrasonography revealed polycystic horseshoe kidney and liver cysts. Computerized tomography showed enlarged kidneys with multiple cysts, fused by their lower poles and multiple liver cysts. She had been hypertensive since her first pregnancy at age 15. Familial history: a brother with horseshoe kidney and three daughters with polycystic kidney. At admission her renal function was noticed to be abnormal (serum creatinine of 2.1 mg/dl; creatinine clearance of 43 ml/min/1.73m²). It developed to advanced chronic kidney disease, undergoing hemodialysis for four years. Nineteen months ago she underwent kidney transplantation with a related living donation (daughter). Immunosuppression with prednisone and tacrolimus. In a follow up, normal renal function and tomography showed a increased in bilaterally with multiple cysts and fusion by the lower poles (Figure 1), liver cysts (Figure 2) and renal graft in the right iliac fossa (Figure 3).

DISCUSSION

Horseshoe kidney is a renal fusion anomaly most common found, with perdition for the male gender and occurring about 0.25% of the population [2,3]. It is usually asymptomatic, but the affected individuals are more prone to a variety of complications such as renal lithiasis, ureteropelvic junction stenosis, renovascular hypertension, polycystic kidneys and polycystic liver [3].

The polycystic kidney disease is an autosomal dominant hereditary disorder: it affects all ethnic groups worldwide with incidences ranging from 1 in 500 to 1 in 5000 births. About 5% of the reported cases can be occurring without previous family history. It has as responsible genes: PKD1 on chromosome 16 and PKD2 on chromosome 4. Widely expressed in extra renal tissues and their encoded proteins, polycystin-1 and polycystin-2,
respectively, are associated to cyst production, cerebral and aortic aneurysms, cardiac valvular abnormality and colonic diverticuli [4,5].

Until our knowledge, we found two cases with a polycystic horseshoe kidney submitted a renal transplant published in the literature. In both cases, a Nephrectomy bilateral before the transplantation was necessary. In our case the Nephrectomy was not necessary [6,7].

REFERENCES


Figure 2 Computerized abdominal tomography showing liver enlarged and multiple cysts (Polycystic liver).

Figure 3 Computerized abdominal tomography showing pelvic kidney graft, without particularities.

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