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Successfull simultaneous pancreas kidney transplantation in a patient with congenital partial lipodystrophy☆

Trasplante simultáneo de riñón y páncreas exitoso en paciente con lipodistrofia parcial congénita

Dear Editor,

Lipodystrophies (LDs) are a heterogeneous group of genetic and acquired disorders that mainly affect women. They are characterised by total or partial loss of subcutaneous adipose tissue (lipoatrophy), which may be associated with accumulation of fat (lipohypertrophy) in different regions of the body. Patients with LDs have extreme insulin resistance, hyperglycaemia, severe hypertriglyceridaemia, low HDL levels and fatty liver. The seriousness of these metabolic complications is correlated with the extent of fat loss.[1,2]

There are 2 hereditary forms:

- Generalised lipodystrophy, which has an autosomal recessive inheritance pattern and is characterised by total loss of subcutaneous adipose tissue, and
- Congenital partial lipodystrophy, the more common form, which has an autosomal dominant inheritance pattern and is linked to partial loss of adipose tissue.[3]

Several researchers have noted that the metabolic abnormalities observed are due to a failure in the functions of regulation of fatty acid storage and release of adipose tissue. There are also genes involved in the effector actions of insulin, and in adipocyte proliferation and differentiation. Different candidate genes have been analysed, such as the insulin receptor gene, the insulin receptor substrate 1 gene and the beta-2 adrenergic receptor gene.[4] The lack of subcutaneous adipose tissue deposits is due to the low concentration of leptin. Hypoleptinaemia alters the hunger/satiety signals in the central nervous system and causes hyperphagia. Then, caloric excess causes fat to accumulate in the muscles and liver and triggers severe insulin resistance, diabetes with high insulin requirements and hypertriglyceridaemia, which creates a high risk of pancreatitis. Patients with LDs also have fatty liver, myocardiopathies and proteinuric nephropathies.[5]

A 41-year-old female patient with a diagnosis of congenital partial lipodystrophy, diabetes diagnosed at age 15, being treated with insulin aspart (115 IU/day), with poor glycaemic control, who had bilateral proliferative retinopathy and end-stage renal disease secondary to membranoproliferative glomerulonephritis type I (by renal punch biopsy [RPB]), in haemodialysis three times per week since April 2012. In addition, she had mixed dyslipidaemia being treated with atorvastatin 20 mg/day and fenofibrate 200 mg/day and HTN. Physical examination (PE): dry weight 66.500 kg; height 170 cm; BMI: 23.01; BP 160/70.

Her limbs looked thin, with loss of subcutaneous fat in the buttocks, thighs and upper limbs. Hypotrophic breasts, slight hepatomegaly, acanthosis nigricans on the neck and hirsutism.

It was decided to place her on the waiting list for a double kidney–pancreas transplant, considering her CKD in dialysis and her diabetes without insulin resistance,

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given her C-peptide level below one and her expected treatment with recombinant human leptin, requested for compassionate use. She received a double kidney–pancreas transplant on 07/09/2015, with an intestinal bypass. She also received induction with ATG-Fresenius (anti-human T-lymphocyte immunoglobulin from rabbits) (15 mg/kg divided into 5 doses), antibacterial prophylaxis for 5 days with piperacillin/tazobactam, an antifungal agent for one month, an anti-cytomegalovirus (CMV) agent (for 3 months) and an anti-pneumocystis pneumonia (PCP) agent for 2 years. Maintenance immunosuppression was added with belatacept (*de novo*) and tacrolimus/Deltisona (methyl-prednisolone)/mycophenolate sodium. The indication for belatacept was due to the patient's vascular/metabolic risk and to initial efforts to decrease the dose of tacrolimus (relative to the standard dose) to later suspend it in the long term. The grafts functioned immediately; neither insulin nor haemodialysis was required. In the postoperative period, the patient had intercurrent intestinal obstruction with an anastomosis fistula, which required conversion to a new Roux-en-Y duodenojejunostomy. She also had a low-output pancreatic fistula, which closed without complications. Currently, the patient has normal renal and pancreatic function, with the following values: fructosamine 237 μmol/l (normal level [NL] 285 μmol/l), HbA1c 4.6% (NL 2.5%–5.9%), C-peptide 1.8 ng/ml (NL 0.5–2 ng/ml), creatinine 0.8 mg/dl (NL 0.8–1.4 mg/dl) and no proteinuria. Tacrolimus levels of 6 ng/ml are being maintained.

This is the first report of a double kidney–pancreas transplant with a favourable course in a patient with congenital partial lipodystrophy and a new immunosuppressant regimen.

Conflicts of interest

The authors declare that they have no conflicts of interest.

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Sjogren syndrome and mixed nephropathy. Significance of early kidney biopsy☆

Síndrome de Sjogren y nefropatía mixta. La importancia de la precocidad en la biopsia renal

Dear Editor,

We present the case of a 76-year-old patient with primary Sjogren's syndrome (pSS) and mild acute renal failure with active sediment. Prior medical history included hypertension well controlled with 2 drugs and pSS with glandular (biopsy) and extraglandular involvement (pulmonary fibrosis, skin purpura and Raynaud's disease that required treatment with steroids), being treated with azathioprine, prednisone 5 mg/day, biphosphonate, calcium/vitamin D,

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