

Table 1. Cases of adrenal insufficiency in haemodialysis patients

Case	52 y/o F	54 y/o F	44 y/o F	33 y/o M	36 y/o F
Type	Primary	Panhypo-pituitarism, primary	Isolated ACTH deficiency	Primary	Primary
Cause	Tuberculosis	Amyloidosis	Unknown	Steroid withdrawal	Steroid withdrawal
Clinical findings	Hyper-pigmentation	Fever	Fever	Fever	Fever
Treatment	PSL 10 mg/day	Cortisone acetate 37.5 mg/day	Without glucocorticoid supplementation	mPSL 30 mg/day	mPSL 20 mg/day
HPA axis	ACTH very high	ACTH not measured	ACTH very low	ACTH not measured	ACTH not measured
Ref.	Neill [1]	Erdkamp [2]	Kato [3]	Sever [4]	Sever [4]

y/o; year-old, F; female, M; male, ACTH; adrenocorticotrophic hormone, PSL; prednisolone, mPSL; methylprednisolone, HPA; hypothalamic-pituitary-adrenal, Ref.; reference.

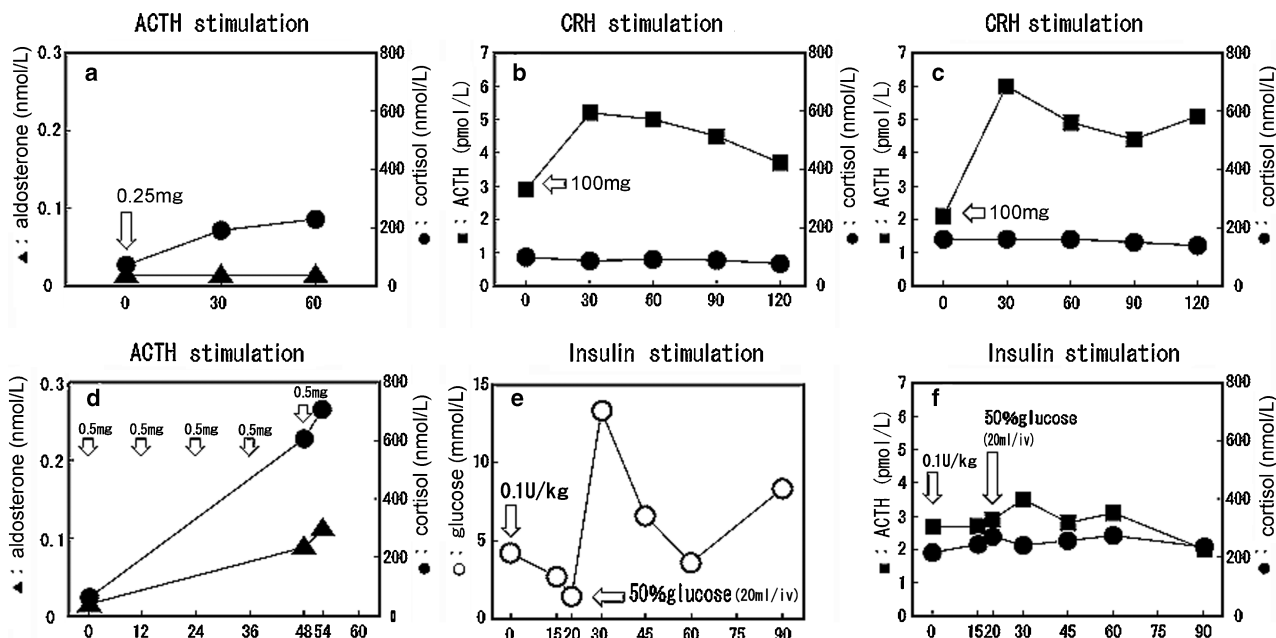


Fig. 1. Time course of stimulation tests to evaluate the HPA axis. Blood levels of aldosterone (▲-▲), cortisol (●-●) and ACTH (■-■) shown, as well as described in the text, were measured at the Health Sciences Research Institute, Yokohama, Japan. Open circles in (e) (○-○) indicate blood glucose levels. Values under a horizontal axis of each figure indicate minutes elapsed. Informed consent was obtained for each test performed.

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Successful treatment of spontaneous kidney graft decapsulation 18 years after renal transplantation: a case report

Sir,

The detection of a perirenal fluid collection causing graft dysfunctions is common in the early postoperative phase after renal transplantation. These are typically lymphocelles, urinomas, abscesses and haematomas, and treatment is well established. Perirenal fluid collection causing acute or sub-acute renal failure several years after engraftment is unusual. A rare, but potentially treatable condition is acute spontaneous decapsulation of the kidney graft. We hereby present a case of spontaneous decapsulation and rapid decline of renal function 18 years after engraftment.

A 41-year-old Caucasian male with end-stage renal failure was diagnosed in 1989 with chronic glomerulonephri-

tis (not biopsy verified) and received a renal transplant in 1990 at age 23, from his HLA-identical 21-year-old brother (CMV+ to +, PRA-). There were no rejections or other early complications. Graft function remained steady over the next 18 years with s-creatinine 100–120 $\mu\text{mol/L}$. Immunosuppression consisted of CsA (125 mg b.i.d., C_0 target level 75–125), prednisolone (5 mg \times 1/day) and azathioprine (75 mg/day, 6-TGN target 100). In 2003, an ultrasound of the renal graft and in 2005 a CT scan of the abdomen was done with normal findings regarding the graft.

During a routine control at the local hospital s-creatinine had increased from a stable value (100–120 $\mu\text{mol/L}$) to 150 $\mu\text{mol/L}$. The development of chronic allograft nephropathy was suspected. Azathioprine was replaced by mycophenolate mofetil (MMF; 750 mg b.i.d.), and an ultrasound-guided renal biopsy was planned. The day after switch from azathioprine to MMF, the patient experienced a sudden onset of severe pain, and swelling in the graft was localized. S-creatinine at admission had further increased to 201 $\mu\text{mol/L}$.



Fig. 1. A CT Scan of the abdomen revealed a fluid collection (A) encasing the renal graft (B).

A CT scan of the abdomen revealed a large fluid collection (>1 L) encasing the renal graft (Figure 1). A pigtail catheter was placed within the cavity. On the first day, 800 ml of yellow-coloured fluid was drained out and analysis showed an albumin content of 9 g/L, creatinine level 102 µmol/L, WBC < 0.1 × 10⁹/L and lactate dehydrogenase 68 U/L. Ultrasound/Doppler examination of the graft (after drainage) demonstrated normal parenchyma, renal pelvis and flow pattern. A renal biopsy was not performed. His symptoms resolved and s-creatinine normalized.

However, fluid secretion through the pigtail catheter persisted, at the level of 100–200 mL/day. A laparoscopic exploration was therefore performed after 1 week. Perioperatively, a huge cyst encapsulating the kidney graft was noted. A fenestration to the peritoneal cavity was easily performed by making a 3-inch window in the anterior/cranial wall of the cyst. The kidney surface was described as intensely red and ‘bare’—well in conjunction with the CT finding of renal decapsulation. Three months after the procedure, the renal allograft was functioning remarkably well with s-creatinine 95 µmol/L.

Discussion

Late spontaneous decapsulation of a kidney graft is a rare event [1–5]. The pathophysiology behind spontaneous decapsulation of kidney transplants remains unclear. Case re-

ports indicate, however, that they all share some common clinical features; the late occurrence (4–12 years post-Tx); subcapsular localization; lymphatic biochemistry; large fluid volume and persisting secretion after drainage.

Several treatment options have been described. Koena *et al.* reported the first two cases in 1979 [1]. They tried to develop a new capsule and drainage by a LeVeen shunt, but failed. Both patients ended up with a graftectomy. In 1982, marsupialization was successfully used for the first time by Nghiem *et al.* [2]. According to Brown *et al.* [3] diuretics solved the problem.

The choice of therapy must depend on the patient’s clinical presentation. In our case, the accumulation of fluid leads to pressure on the kidney and rapid deterioration of graft function. The biochemical composition of the fluid indicated lymphatic or transudative character.

He was switched from azathioprine to mycophenolate mofetil, but that was after s-creatinine had started to rise. To our knowledge, this is the latest post-Tx time point for spontaneous decapsulation reported in the literature. This is a rare complication, but with a deleterious effect on kidney function, if not detected and treated adequately. The exact cause of the late, spontaneous decapsulation remains obscure. But the incident seems to be inherent to transplanted kidneys, and the progressive tissue weakening possibly caused by the immunosuppressive therapy could be a contributing factor for the tissue disruption.

Conflict of interest statement. None declared.

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IgG4⁺MOLPS associated with inflammatory abdominal aortic aneurysm, interstitial pneumonia and interstitial nephritis

Sir,

A 74-year-old man presented with anorexia, general fatigue, weight loss and purpura on the lower legs. Auscultation revealed trivial fine crackle. The white blood cell count was 5600/mm³, with 6.0% eosinophils. Haemoglobin

was 8.5 g/dl. The C-reactive protein was 5.2 mg/dl. BUN, creatinine and uric acid were 11, 0.86 and 5.9 mg/dl, respectively. Liver enzymes were within normal limits. IgG, IgA and IgM were 6170 mg/dl, 129 mg/dl and 38 mg/dl, respectively. IgG1, IgG2, IgG3 and IgG4 were 3470, 1460, 552 and 807 mg/dl. IgE was 1070 IU/ml. Proteinuria was 100 mg/dl (0.7 g/day), and trivial microhaematuria was observed. NAG was 23.1 U/g-Crn. CH50, C3 and C4 were <6.3 CH50/ml, 19.2 mg/dl and <2.0 mg/dl, respectively. CIq was 36.0 µg/ml. The antinuclear antibody was 1280×. Rheumatoid factor and anti-cardiolipin β2 glycoprotein-I antibody were positive. Other serological tests, including ANCA, anti-ds-DNA antibody, anti-Sm antibody and anti-RNP antibody, were all negative. Enhanced computed tomography (enhanced CT) detected interstitial pneumonia and abdominal aortic aneurysm (AAA) with a wall thickness measuring 3.2 × 3.0 cm (Figure 1A). ¹⁸F-fluorodeoxyglucose positron emission tomography-CT (¹⁸F-FDG PET/CT) demonstrated increased

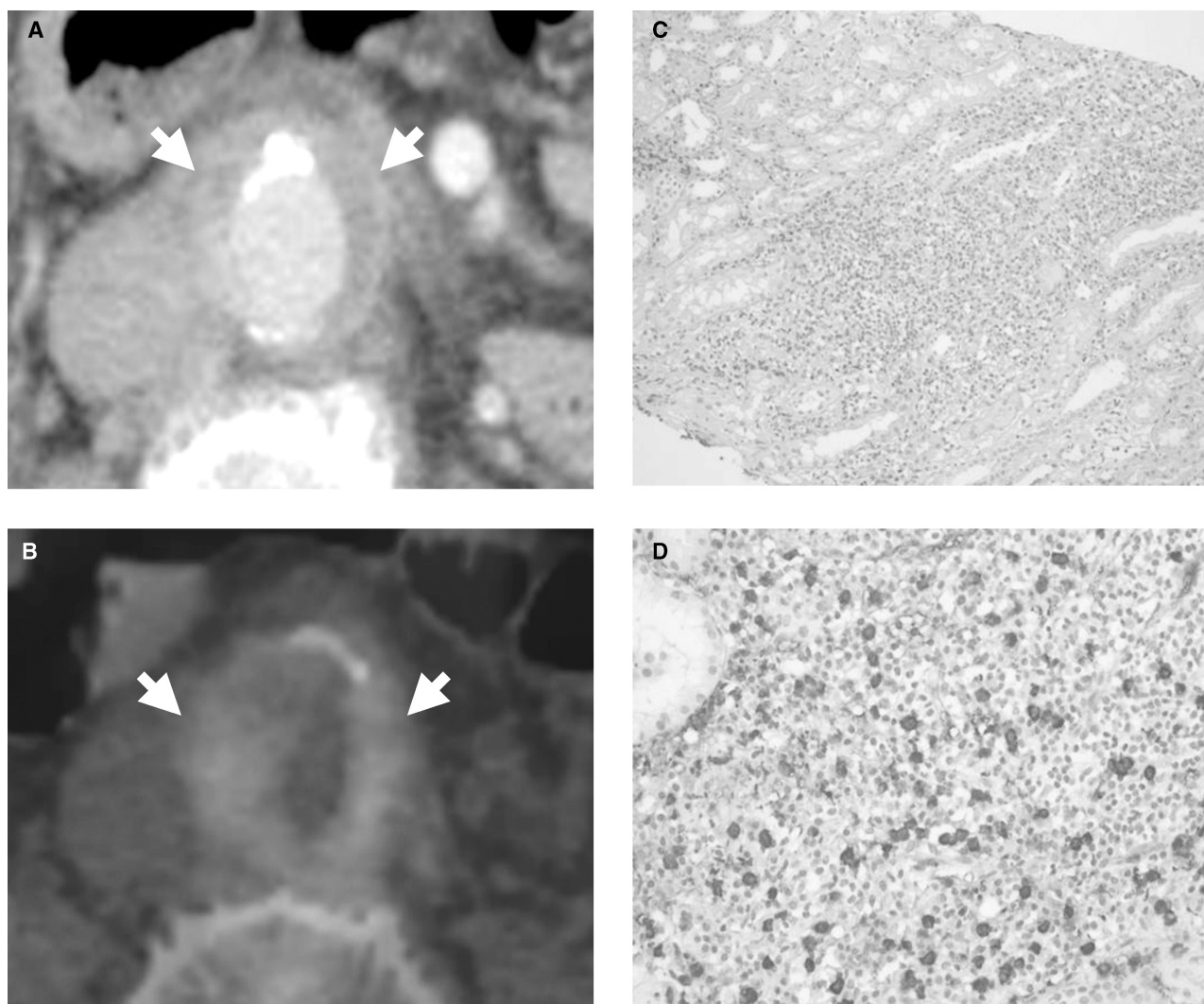


Fig. 1. Findings of enhanced CT, ¹⁸F-FDG PET/CT and renal biopsy. (A) Enhanced CT reveals wall thickness of abdominal aorta, compatible with IAAA (arrow). (B) PET-CT detects FDG uptake within thickened aortic wall, compatible with IAAA (arrow). (C) Light microscopic findings of a renal biopsy specimen. Patchy patterns of plasma cells infiltrations are observed in the interstitium. PAS stain. 200×. (D) Immunostaining of anti-IgG4 antibodies detected many IgG4⁺ plasma cells in the interstitium.