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C. Cobelo, E. Sola, V. López,
C. Gutiérrez-de la Fuente,
D. Hernández Marrero
Nephrology Department. Carlos Haya Regional Hospital. Málaga, Spain
Correspondence: C. Cobelo
Servicio de Nefrología. Hospital Regional Universitario Carlos Haya. Málaga.
cofecamu@hotmail.com

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Hyponatraemia, Hypopotassaemia and Pre-renal Acute Renal Failure as a Presentation of Cystic Fibrosis

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Dear Editor,

Cystic fibrosis is a hereditary disease with a recessive autosomic pattern. It is characterised by ecrine and exocrine gland abnormal function that causes chronic lung conditions, abnormally concentrated sweat and pancreatic failure. The usual symptoms are seen in infancy and are respiratory, such as cough and recurrent respiratory and digestive complications such as meconium ileus.[1] A reduced percentage of cases reaches adulthood without being diagnosed due to partial mutations.[2] It is rarely associated with volume decrease and metabolic alkalosis and it is seen during the summer season due to lack of fluid loss replenishment.

We present the case of an adult man who entered the nephrology department with a diagnosis of volume decrease, alkalosis, hypopotassaemia and acute renal failure.

Case report

The case is a 29 year old man who had been referred 6 years before to nephrology outpatient services by the emergency room because he was suffering from intense diaphoresis, muscular weakness and arthralgia. In the emergency room the following laboratory values had been found: Na 128, K 2.8, urea 71 and glucose 139.

Among his personal medical history were multiple fractures due to a traffic accident, appendectomy and frequent respiratory infections. On physical examination he presented a blood pressure of 110/74 mmHg, a heart rate of 100 bpm and skin and mucose membrane dehydration. Nothing else of note was seen in the course of the physical examination. The most relevant values seen in the laboratory were: (emergency room analysis) Hb 17.8, Na 128, K 2.9, urea 71, pH 7.56, HCO_3 25, PCO_2 28.9, base excess 3.30; (consultation analysis: 2 days later, when the emergency room had already rehydrated the patient) Hb 13.6, VSG 6, Cr 0.87, lipase 1,287, osmolarity in serum 2.94, Ca 8.87, phosphatase 2.6, pH 7.35, PCO_2 57, HCO_3 31.2, Na(O) 18 mEq/24 h, Cl (O) 8 mEq/24 h, proteinuria 0.21 g/24 hours. The condition was interpreted as heat stroke.

The following year, coinciding with the summer season, the patient once more had intense diaphoresis, renal failure, hypopotassaemia and hyponatraemia that were resolved with hydration. The patient was advised to ingest fluids and electrolytes during the summer.

The following year the patient suffered from the same condition and in view of the history of profuse sweating as a consequence of the hydroelectrolyte disorder, a sweat test was requested and they found positive the following: chlorine (sweat) 83 (normal 28). A genetic test for cystic fibrosis using the INNO-LIPA CFTR 12 and CTFR 17 technique was normal.

The next year the patient had a recurrence of the same clinical condition, and suspecting cystic fibrosis, a spermogram was performed and showed complete azoospermia. On abdominal CT a pancreas with thickened body and head was seen. There were no other findings. The genetic study using the OLA-PCR technique was repeated and the results were normal, although CFTR (IVS8) 5T polymorphism was found. Since further abnormalities were suspected the molecular study of the CFTR gene was continued. For this reason, a genetic-molecular study was requested and the following genotype obtained: 1811+1,6kba>G/A1006E, 5T,V562I. The patient was sent to the cystic fibrosis unit.

Discussion

The relation between salty skin and early death has been known for a long time, cystic fibrosis was described in 1930 and was called cystic fibrosis of the pancreas because this organ was the most affected, even though later it was known that the condition affected all exocrine glands.[3] The disease is due to the mutation of the cystic fibrosis gene that codes for a membrane protein that can be a ion canal, this causes an alteration of chloride secretion and modifies all exocrine secretions.

The case is a young patient who comes to consultation due to dehydration in summer. Once the patient recovered, no further studies were performed, but when the patient had a recurrence of symptoms during the same time of year further attention was given to the sweating as it was considered related to the reason for the condition. The sweat test is the most important diagnostic method for the detection of cystic fibrosis.[4] And in this case it was positive. Subsequently a genetic test was requested, this was negative and once the patient recovered, there was no further insistence on

achieving a diagnosis. When the clinical condition recurred, a spermogram was requested, since more than 97% of patients with cystic fibrosis are sterile due to incomplete development of the wolffian ducts. Absolute azoospermia was found. A first genetic analysis was negative, the second (covers 76% of the Spanish population) analysis only found a polymorphism, and the study was continued. The last test (covers 96% of the Spanish population) showed two mutations of the CFTR gene (1811+1,6kba>G/A1006E, 5T,V562l).

The diagnosis of cystic fibrosis was carried out by means of a positive sweat test and tests that showed two gene mutations.[5-7]

Hypovolaemia with recurrent metabolic alkalosis during the summer must lead us to suspect cystic fibrosis as a possible cause.

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M.A. Pavone, A. Solís Padrones, D.G. Muratore, M. Saiz, C. Puig
Nephrology Department.
Hospital Can Misses. Ibiza, Balearic Islands, Spain.
Correspondence: Mario Alberto Pavone
Servicio de Nefrología. Hospital Can Misses. Ibiza. Islas Baleares. Tel: 971397000
mpavone@asef.es

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Acute Failure of Renal Graft due to *de novo* AA Amyloidosis in a Patient Affected by Gangrenous Pyoderma

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Dear Editor,

Secondary systemic amyloidosis (AA) is a frequent condition, associated with long evolution inflammatory and infectious diseases, as also with some neoplasias. During the first 15-20 years, the disease presents no symptoms, and usually renal involvement is the first clinical sign to appear and manifests itself in the form of proteinuria. It usually evolves to terminal renal failure 2-10 years later.[1] Furthermore, amyloidosis relapse during kidney transplant is also a widely described condition.[2] Primary (AL) amyloidosis or *de novo* secondary (AA) amyloidosis during renal transplant, are considered slowly progressive diseases, which in most cases are not associated with graft loss[3] or only cause graft loss after years of evolution.[4]

Gangrenous pyoderma (GP) is a well-defined skin clinical pathological condition, characterised by the presence of single or multiple erythematous pustules that rapidly progress to necrotic ulcers. Its cause is unknown, although some defects of the oxygen metabolism of neutrophils, overexpression of cytoquines (interleukin-8, interleukin-16) and anomalies of humoural and cell immunity have been described as possible causes, but none is specific.[5] In mild forms, treatment with topical or intralesional steroids is used, but it is always insufficient, and systemic treatment is necessary. Cases that are refractory to steroids can respond to other immunosuppressants, such as oral cyclosporin and sometimes mofetil mycophenolate, azathioprine or methotrexate are also effective, as also new biological therapies (anti-TNF monoclonal antibodies).

We present the case of a 38 year old woman, with a history of well controlled hypertension of long evolution and GP of the lower limbs diagnosed in 1989. Her GP was treated with calcineurin inhibitors (cycosporin and tacrolimus), without any response. She presented multiple infectious complications due to multi-resistant germs that made prolonged antibiotic therapy necessary. Monitored in our service for chronic kidney disease (CKD), a renal biopsy was performed in February 2002, and the report informed of the presence of sclerosis and patchy glomerular atrophy, with vascular hyalinosis, compatible with anticalcineurin treatment side effects.

In August 2002, the patient began substitute renal therapy with haemodialysis, and in October 2005 received a kidney transplant from a deceased donor. On discharge, plasma creatinine levels were 1 mg/dl without proteinuria. For 4 years she received therapy with tacrolimus, mofetil mycophenolate and prednisone at low doses. Renal function was stable, with plasma creatinine of around 1.1 mg/dl and proteinuria kept at <0.5 g/24 hours in spite of presenting several complications in the form of secondary infections to GP. In February 2009 renal function deteriorated (plasma creatinine 1.5 mg/dl) and there was also an increase in proteinuria (2.8 g/24 hours) (Table 1). The possible differential diagnoses were: chronic graft nephropathy, toxicity due to calcineurin drugs, *de novo* glomerular disease, tubule-interstitial or obstructive nephritis. On physical exam the patient was normotense and presented decrease of adipose tissue related to long term protein-calorie malnutrition, lesions in lower limbs secondary to GP and mild discomfort on palpation of the renal graft area. A complete laboratory profile was obtained for glomerular diseases, including a proteinogram and electroimmunophoresis in blood and urine; all tests were negative.

On kidney ultrasound a graft measuring 13.2 x 6.2 cm was detected, with preserved parenchyma and echostructure with no obstructive evidence. On 23rd May 2009 a renal