matrix in 3, periglomerular fibrosis in 2 and negative direct immunofluorescence in 2 with deposits of IgG in 1, C'3 in 4, IgM in 4 and C'1q in 1 case located in the mesangium and subendothelium. EM was only performed in 2 cases, both revealed podocyte fusion and lack of immune complex. In just one[2] of the cases described, renal interstitial granulomas were found with 12.5% involvement, lower than that reported in literature (15-40%). The good evolution of GFR and proteinuria with conservative treatment was notable and it did not improve by adding corticosteroids for their lung disease.

This association can be paradoxical. In sarcoidosis, there is an immune imbalance with upregulation of the TH1 pathway, and in FSGN upregulation of the TH2 pathway.[8,9] However, in late stages TH17 predominates, inducing secretion of interleukin 17[10] favouring the development of fibrosis. Cardiotrophin-like cytokine 1, involved in the development of FSGN, is increased in pathway TH17,[11-13] which may be the link between both.[11]

We present the case of a patient with FSGN onset who that year was diagnosed with sarcoidosis, with good pulmonary and renal evolution on treatment with corticosteroids and blockade of the renin–angiotensin-aldosterone system.

Conflicts of interest

The authors declare that they have no conflicts of interest related to the contents of this article.

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Long-term renal prognosis of typical haemolytic-uraemic syndrome suffered in infancy

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To the Editor:

Renal damage suffered in infancy in the haemolytic-uraemic syndrome (HUS) may determine long-term renal prognosis. We report a relevant case.

Our patient is a 33-year-old woman. When she was 9 months old, she suffered typical HUS that required acute peritoneal dialysis for 15 days, resulting in subsequent full recovery of renal function. She was discharged from consultations when she was 16 years of age. Since then, she has had no other relevant illness. Normotensive. She sought consultation due to progressive proteinuria with 5 years of evolution was determined by urine test strip. She is a woman of normal size, height 167cm, weight 67kg, blood pressure: 123/80. No oedema. The remainder was without findings. In complementary tests: CRP was 0.89mg/dl, creatinine clearance was 87ml/min, non-selective glomerular proteinuria was 292mg/dl (2.29g/d) urine analysis with absence of microhaematuria. Cholesterol was 252mg/dl. Total protein was 6.8g/d, albu-

minaemia was 4.0g/d. Complete blood count and angiotensin converting enzyme were normal. Immunology, thyroid profile and basic viral serology were negative. Renal ultrasound with kidneys showed a diameter of 97 and 95mm, with cortex of 13mm and 10mm and resistance index of 0.6 and 0.65. Immunoelectrophoresis of proteins in blood and urine and normal light chains. Computerised tomography of the chest, abdomen and pelvis without findings. Mantoux test negative. Renal biopsy was performed, revealing 7 glomeruli: 1 sclerotic, 4 normal and 2 with signs of focal segmental hyalinosis (Figure 1). Immunofluorescence: negative. Electron microscopy: not performed. Treatment was started with angiotensin converting enzyme inhibitors with reduction in proteinuria at 3 months to 1 g/d and glomerular filtration stability.

In childhood HUS, the number of affected capillaries determines long-term glomerular damage by developing glomerular hyperfiltration (GHF). Many articles[1-5] recognise this fact. Garg et al.[6] include 49 studies, 3476 patients with a mean follow-up period of 4.4 years (1 to 22), patients from 1 month to 18 years old. It shows a combined incidence of mortality/end-stage renal failure (ESRF) of 12%. 64% displayed renal complications (defined as high blood pressure [HBP], a decrease in the glomerular filtration rate [GFR] <80ml/min and/or significant proteinuria), with a combined incidence of 25%. 15% had proteinuria, 10% HBP and there was a 15.8% drop in GFR. The long-term prognosis was worse in those who suffered cortical necrosis and who required renal replacement therapy for longer than 8 days. Between 8% and 61% of those who had fully recovered renal function suffered renal complications that even began 20 years later.

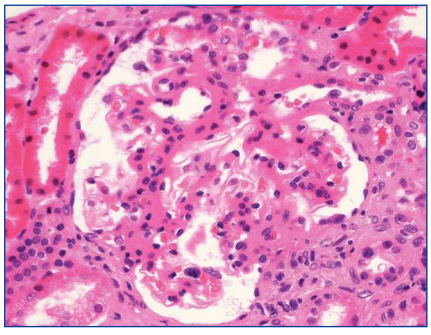

Figure 1. Kidney biopsy.
Optical microscope (haematoxylin and eosin technique) (400x): focal and segmental hyalinosis.

Moghal et al.[7] biopsied 7 normotensive patients with complete recovery of renal function and late damage, with the following result: overall glomerulosclerosis (85.7%), segmental sclerosis lesions (28.6%), tubular atrophy (14%) and overall glomerulomegaly with intimal thickening of small vessels (100%).

Caletti et al.[8] biopsied 30 children with renal complications with 11.2 years evolution. They found 56.6% of focal segmental glomerulosclerosis with hyalinosis, 30% of diffuse mesangial proliferative glomerulonephritis (GN), 6.6% of GN with minimal changes and 6.6% of diffuse glomerulosclerosis. The findings were interpreted as mesangial GN followed by focal segmental glomerulosclerosis with hyalinosis culminating in diffuse glomerulosclerosis. After follow-up, only 25% of the latter had a normal GFR.

Tönshoff et al.[9] studied 89 patients after 16 years. 10.4% progressed to advanced chronic renal failure (CRF) and 3% to ESRF.

Gagnadoux[4] followed up 29 patients for 15-28 years. 41.4% had sequelae, 10.3% progressed to CRF and 13.8% to TRF between 16 and 24 years after it started. 6.9% had normal GF at 10 years and developed CRF after this time.

Kelles et al.[5] followed up 95 patients for 10 years and found that 65% did not suffer sequelae, 26% had mild renal disorder and 9% had progressed to severe CKD.

In all series, the possibility of progression was greater if they suffered mesangial GN.

These data coincide with the possibility of developing late renal failure after suffering pre-eclampsia in pregnancy.[10] In both cases, a noxa, self-limiting in time determines functional renal sequelae in the very long-term.

In our case, the histology ruled out the possibility of primary glomerulopathy. In spite of the short follow-up time, we found a significant decrease in proteinuria, giving the patient a good renal function prognosis.

This case illustrates that a typical epidemic HUS in children may cause renal failure, HBP and proteinuria. These complications may even begin 20 years after the illness has been considered cured. As such, we indicate long-term follow-up of these patients. Treatment should be aimed at controlling risk factors that accentuate GHF (obesity, HBP) and specifically inhibition of the renin-angiotensin-aldosterone axis.

Conflicts of interest

The authors declare that they have no conflicts of interest related to the contents of this article.

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An uncommon cause of high blood pressure in young people: retroperitoneal paraganglioma with vascular invasion

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To the Editor:

Paragangliomas are rare extra-adrenal tumours that emerge from the autonomic nervous system. They are classified as functioning or non-functioning tumours, depending on the production of catecholamines. It presents clinically as high blood pressure associated with vegetative symptoms.

CLINICAL CASE

We report the case of a 20-year-old male who presented with hypertensive emergency that had begun a year before, accompanied by profuse sweating, generalised flushing, headaches and occasionally, lower back pain. Blood pressure reached 230/110mmHg, often triggered by physical effort. A physical examination revealed malar rash and palmar erythema that disappeared with acupressure. Laboratory tests showed an increase in catecholamines and metanephrines in urine at 24 hours: noradrenaline excretion of 7515.77nmol/24h (<504), dopamine of 4298.84nmol/24h (<3237) and normetanephrine of 39676nmol/24h (<2424).

An abdominal ultrasound revealed a retroperitoneal mass of about 8cm of heterogeneous echogenicity and marked vascularisation. Given these findings, a computerised axial tomography (CAT) scan of the abdomen and pelvis was performed that confirmed the existence of a retroperitoneal mass of 8 x 4cm above the aortic bifurcation that infiltrated the inferior vena cava and bordered 180° of the aorta, covering the inferior mesenteric artery (Figure 1). Given the findings of the CAT scan, a cavography was performed to evaluate resectability, revealing an infrarenal intracaval filling defect 2cm in diameter (Figure 2).

Suspecting paraganglioma of the organ of Zuckerkandl, beta-adrenergic blockade was produced and we subsequently performed a surgical en bloc resection of the tumour; we partially sectioned the anterior vena cava to remove the intracaval tumour. The postoperative course was uneventful, the symptoms disappeared and plasma and urine catecholamine and metanephrine levels normalised.

The anatomopathological diagnosis confirmed the suspicion of well-delimited and encapsulated paraganglioma, with a low proliferative index (Ki 67: 5-7%). The patient was referred to the Medical Oncology department, where a 123I-MIBG scintigraphy was requested that showed no signs of residual or distant disease. In the absence of data that justified adjuvant chemotherapy, it was decided to follow up the patient periodically.

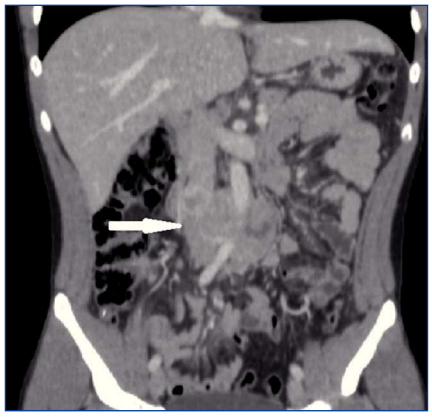

Figure 1. Tomografía computarizada abdómino-pélvica (corte sagital). Tumoración retroperitoneal lobulada, heterogénea, que bordea la aorta e infiltra la vena cava inferior.