diameter. When we were monitoring the patient's CKD in our department, the patient also presented persistent hypokalaemia due to hyperreninemic hyperaldosteronism secondary to the underlying disease (malignant HT), which ruled out the possibility of a functional adrenal mass. However, as part of the pre-transplant study in 2008, the patient underwent a CT-guided needle biopsy of the adrenal mass, and the results from the histological study suggested a myelolipoma, thus confirming the initial diagnosis. As the tumour was benign, it did not contraindicate kidney transplantation.

During the two-year outpatient monitoring period following the transplant, the patient presented refractory HT requiring six different hypotensive drugs to achieve rather poor blood pressure control. His renal function deteriorated slowly over this time, and he presented proteinuria and microhaematuria. The persistent hypokalaemia reappeared and doctors ordered a new hormonal study. This time, the study found high plasma aldosterone (1098pg/ml) and suppressed plasma renin activity (0.13ng/ml/h). The patient was then diagnosed with primary hyperaldosteronism and the CT and MRI scans were repeated; the adrenal mass had reached 12*5cm in diameter along the cranio-caudal plane and 10cm in diameter along the transversal plane. It contained mainly fatty tissue with dense soft tissue areas. The patient was referred to the General Surgery Department, and in March 2010, underwent laparoscopic right adrenalectomy with excellent and prompt recovery.

The histological study showed an adrenal myelolipoma with hyperplasia of the adrenal cortex (zona glomerulosa) secondary to the pressure exerted by the large size of the myelolipoma. This explained the patient's primary hyperaldosteronism, even though the tumour was benign and non-functional.

We could reduce the hypotensive drugs by half in the post-operative phase. The patient now has excellent control over his hypertension with the aid of two hypotensive drugs and blood potassium levels are normal, which suggests that the renal hyperplasia was not bilateral and was clearly associated with the myelolipoma.

We found cases of myelolipomas associated with arterial HT in the literature, but the tumours have never been shown to be functional. Arterial HT was rather explained by renovascular causes, due to pressure exerted by the tumour, or associated with obesity or endocrine conditions such as Cushing's syndrome or Conn's syndrome. This case is exceptional as primary hyperaldosteronism was caused by a myelolipoma, which could possibly be explained by the pressure exerted on the adrenal gland by the large tumour.

Conflicts of interest

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

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Reactive haemophagocytic syndrome associated with parvovirus B19 in a kidney-pancreas transplant patient

Nefrologia 2012;32(1):125-6
doi:10.3265/Nefrologia.pre2011.Oct.11179

To the Editor,

Reactive haemophagocytic syndrome or secondary haemophagocytic lymphohistiocytosis (HLH) is a disorder of the mononuclear phagocyte system characterised by generalised, ineffective and uncontrolled histiocytic proliferation that leads to cell damage and multiple organ dysfunction with haemophagocytosis. The first description of secondary forms of this disease was by Risdall et al,[1] who in 1979 described a syndrome characterised by a proliferation of histiocytes with haemophagocytic activity, associated with a viral infection. This syndrome was later described in association with infections of all types and with non-infectious diseases such as rheumatoid arthritis, lupus, leukaemia, lymphomas, myelodysplastic syndromes and carcinomas.

Its pathogenesis is still unclear, although there are several hypotheses. The development of this syndrome is likely to be due to an immunological disorder that results in uncontrolled T-lymphocyte activation,[2] causing hypercytokinaemia, and consequently, excessive macrophage activation.

It is diagnosed according to the criteria in HLH-2004[3] and the treatment focuses on the infectious process, as well as on the use of gamma globulin and immunosuppression.[4]

We describe the case of a patient aged 42 years with a history of type 1 diabetes, diabetic nephropathy, and chronic renal failure who underwent a kidney-pancreas transplant (October 2010). The patient's maintenance immunosuppressants are deltisone B, everolimus, and tacrolimus (FK) in addition to prophylaxis with valgancyclovir and trimethoprim/sulfamethoxazole (TMS). The patient experienced fever, vomiting and odynophagia for 2 weeks, and was treated with oral antibiotics. Fever, asthenia and dehydration persisted, so the patient was hospitalised. Laboratory analyses revealed pancytopoenia and renal and pancreatic dysfunction; the patient received subcutaneous insulin but not haemodialysis. Blood and urine cultures were performed, as well as a PCR (polymerase chain reaction) test for cytomegalovirus (CMV), and empirical treatment with ceftriaxone and ciprofloxacin was administered. Twenty-four hours after admission, the haemodynamic state had deteriorated severely and the patient was moved to intensive care, where all immunosuppressants except for corticosteroids were discontinued. Antibiotic coverage was increased through vancomycin, imipenem, fluconazole and ganciclovir. Seven days after admission, the patient was still feverish with positive cultures for common microbes and fungi and a negative PCR for CMV. We ordered PCR for parvovirus B19 due to the persistent pancytopoenia. The physical examination showed cutaneous and mucosal pallor, asthenia, adynamia and splenomegaly. The laboratory results were as follows: Hb: 8.6mg/dl, leukocytes: 900mm^3, triglycerides: 317mg/dl; ferritin >1500mcg/l. In light of suspected haemophagocytic syndrome (5 criteria met), we performed a bone marrow biopsy, which revealed histiocytes with haemophagocytosis. The patient was treated with high doses of gamma globulin (400mg/kg) during 5 days. In the end, PCR was positive for parvovirus B19.

All of the patient's low values improved (haematocrit 28%; Hb 9.4g/dl; leukocytes 1900mm^3; platelets 203 000mm^3) and immunosuppressant treatment was resumed. Pancreatic function remained weak, and the nephrology department found the renal function to be so severely affected that the patient needed haemodialysis. A kidney biopsy puncture was performed which yielded insufficient material.

At 30 days of hospitalisation, the patient was once again feverish with a headache; lumbar puncture revealed normal cerebrospinal fluid, acid-alcohol resistant bacilli (AARB) negative; PCR for CMV, herpes simplex virus, Epstein-Barr virus, cryptococcal antigenaemia all negative; adenosine deaminase at the upper cut-off level; cerebral MRI showed no lesions. The thoracic radiography showed bilateral interstitial and alveolar infiltrates, which was confirmed by thoracic CT as bilateral radiodense infiltrates; fibrobronchoscopy with bronchoalveolar lavage was performed; negative for AARB and positive for pneumocystitis carinii (PCP) when TMS treatment began. Due to the persistent fever and the lack of culture isolation in a case with pulmonary lesions, empirical treatment with isoniazid, rifampicin, ethambutol and liposomal amphotericin was administered. Another kidney biopsy puncture was performed, but graft bleeding ensued and the patient had to go to the surgical ward. Doctors decided to extirpate both grafts, and observed mesenteric adenopathies and abundant purulent matter. This matter tested AARB (+) under direct examination, and therefore antibiotic and antifungal treatments were suspended, with the patient continuing tuberculosis treatment. Final culture was positive for tuberculosis. Patient's fever subsided and overall condition improved; he returned to his home city and is monitored by his local haemodialysis centre.

Haemophagocytic syndrome that reacts to associated infections is a severe and potentially fatal condition. Immunosuppressed patients who present with a fever and haematological abnormalities (cytopoenias) should be screened for haemophagocytosis as early diagnosis enables proper treatment and a favourable prognosis.

Conflicts of interest

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

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