Kimura’s disease in a patient with idiopathic dilated cardiomyopathy on ambulatory peritoneal dialysis

Sir,

Kimura’s disease, endemic in Asia, manifests as solitary or multiple subcutaneous nodules in the head and neck, marked peripheral eosinophilia and elevated serum IgE [1]. Many cases of this are associated with nephrotic syndrome of unknown cause; and there is no evidence in these patients for a relationship between the end-stage renal disease and Kimura’s disease. We describe a patient on chronic ambulatory peritoneal dialysis (CAPD) with a unique concomitant presentation of idiopathic dilated cardiomyopathy and Kimura’s disease.

A 17-year-old Taiwanese boy was diagnosed as having nephrotic syndrome when he was 12 years old, and he was treated with oral prednisolone. Then he was lost to follow-up, having turned to traditional Chinese herbal drugs. At the age of 16, he presented at our hospital again with dyspnoea and oliguria. Haemodialysis was performed initially because of acute pulmonary oedema; then he received CAPD. Idiopathic eosinophilia was noted at this admission [white blood cells (WBCs), 6050/μl; segmented neutrophils, 59.4%; lymphocytes, 10.9%; monocytes, 4.1%; eosinophils, 25.3%; basophils, 0.3%; absolute eosinophil count, 1530/μl (normal 50–450/μl)]. His IgE was 2098 IU/ml (normal < 200 IU/ml) and his haemoglobin was 9.9 mg/dl. About 6 months later, two subcutaneous nodules were found in his neck. Excisional biopsy was done which showed florid germinal centres, eosinophilic infiltration, eosinophilic abscesses and an increase of post-capillary venules—findings compatible with Kimura’s disease [2]. The patient’s cardiothoracic ratio (CTR) was 57.8%. Cardio-echography revealed dilatation of all four chambers with poor left ventricular performance [left ventricular ejection fraction (LVEF) 46%] and moderate pericardial effusion. Idiopathic dilated cardiomyopathy was diagnosed. The patient, however, refused pericardiocentesis or myocardial biopsy. We tried methylprednisolone 500 mg intravenously for 3 days then oral prednisolone 40 mg/day tapered to 10 mg/day for 6 months. His eosinophil count decreased from 3429 to 391/μl (from 34.5 to 6.1 as a percentage of WBCs). His CTR remained essentially unchanged (58.2%), and his left ventricular function did not improve (LVEF 45%) after 6 months of steroid treatment. Since then, the patient has not received steroids, because he has had no appreciable improvement of cardiac function.

A systemic component is usually absent in Kimura’s disease, but the disease is often associated with proteinuria and nephrotic syndrome [3]. In our case, we could not identify the aetiology of idiopathic dilated cardiomyopathy because myocardial biopsy was not performed. We can, however, exclude dialysis-related cardiomyopathy, because the patient was well nourished and free from oedema, and he had the following laboratory parameters: creatinine clearance (Ccr), 73.821/week; weekly Kt/V, 2.46; and normalized protein catabolic rate (nPCR), 1.59 g/day. Based on a report by Sekiguchi et al., that steroid therapy was effective in 16 of 25 cases of eosinophilic heart disease [4], we tried steroids on this patient for 6 months, at the end of which his eosinophil count was decreased but without significant improvement of his left ventricular function.

In conclusion, this is a unique case in which Kimura’s disease and idiopathic dilated cardiomyopathy co-existed. Their as yet undefined relationship may be evaluated further.

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Gas-forming infection in a renal cyst of a patient with autosomal dominant polycystic kidney disease

Sir,

Autosomal dominant polycystic kidney disease (ADPKD) often leads to progressive renal failure due to enlargement of the cysts. Other renal manifestations that can occur include hypertension, urinary tract infection, haematuria, nephrolithiasis, and acute or chronic flank and abdominal pain. Renal cell carcinoma is an infrequent complication of ADPKD [1]. Approximately 30–50% of patients with ADPKD will have one or more renal infection during their lifetime. The primary clinical manifestations of renal infection in polycystic kidney disease are fever and flank pain, which may be associated with bacteremia [2]. These infections may be due either to parenchymal infection or to an infected cyst. Distinguishing between these disorders is often difficult [3]. We present a case of adult polycystic kidney disease complicated by infection of the cysts with gas formation.

Case
A 40-year-old male patient was diagnosed with adult polycystic kidney disease with chronic renal failure on treatment with haemodialysis. The patient was admitted to the Nephrology department with fever and right flank pain. His laboratory findings were as follows: creatinine 11.5 mg/dl, urea 169 mg/dl, alanine aminotransferase (ALT) 17 U/l, aspartate aminotransferase (AST) 112 U/l, albumin 2.6 g/dl,
lactate dehydrogenase (LDH) 481 U/l, leukocytes 9200, haemoglobin 7.2 g/dl, C-reactive protein (CRP) 187 mg/l. Abdomen radiograph showed an air level in the right upper abdominal quadrant (Figure 1). Abdominal ultrasonography confirmed the gas within the cyst in the right kidney. Abdominal computed tomography (CT) scan demonstrated the presence of gas within the cysts (Figure 2). Antibiotics were started (metronidazole, quinolone). Urgent nephrectomy was planned, but the patient died with sepsis.

Infection of cysts in adult polycystic kidney disease is rare. In a few cases, cyst infection with gas was reported. Zijl et al. reported an emphysematous cyst in the right native kidney which 7 months previously had been a haemorrhagic cyst in a kidney transplanted patient with ADPKD [4]. Lanzas et al. presented a haemodialysis patient with adult polycystic kidney disease who presented with sepsis secondary to infection of the renal cyst with gas formation [5]. These two cases were successfully treated with urgent nephrectomy. To our knowledge, this is the third case of cyst infection with gas formation in an ADPKD patient reported in the literature.

Infection of cysts with gas formation in end-stage renal failure patients due to adult polycystic kidney disease is a very rare entity and urgent nephrectomy may save their lives.

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