Small intestinal infarction: a fatal complication of systemic oxalosis

J S Johnson, A K Short, A Hutchison, N R Parrott, I S D Roberts

Abstract

Primary hyperoxaluria is a rare genetic disorder characterised by calcium oxalate nephrolithiasis and nephrocalcinosis leading to renal failure, often with extra-renal oxalate deposition (systemic oxalosis). Although ischaemic complications of crystal deposition in vessel walls are well recognised clinically, these usually take the form of peripheral limb or cutaneous ischaemia. This paper documents the first reported case of fatal intestinal infarction in a 49 year old woman with systemic oxalosis and advocates its consideration in the differential diagnosis of an acute abdomen in such patients.

(Keywords: primary hyperoxaluria; oxalosis; intestinal infarction)

Case report

A 49 year old white woman developed rapidly progressive renal failure, diagnosed two months after a total abdominal hysterectomy and bilateral salpingo-oophorectomy for fibroids, requiring haemodialysis four months later. In her past medical history she had two episodes of renal calculi at the ages of 4 and 24 years, which required surgery but were not investigated further. Before the hysterectomy, renal function was normal and histology of the hysterectomy specimen showed no evidence of crystal deposition. Ultrasound scan excluded urinary tract obstruction. A renal biopsy performed after her stabilisation on haemodialysis showed massive oxalate crystal deposition within tubules, with associated tubular obstruction and acute on chronic tubular injury. A clinicopathological diagnosis of primary hyperoxaluria was made. She was subsequently started on pyridoxine and citrate and consequently decreased transamination of glyoxylate aminotransferase (AGT). The consequent decreased transamination of glyoxylate to glycine leads to a subsequent increase in its oxidation to oxalate, which is a poorly soluble end product. The disorder is characterised by recurrent calcium oxalate nephrolithiasis and nephrocalcinosis, frequently leading to renal failure and death before the age of 20 years. Although more common in early childhood, the age at onset of symptoms ranges from birth to the 6th decade. The disease is classified into two types, type I (glycolic aciduria) and type II (L glycric aciduria), on the basis of the different patterns of urinary organic acid secretion.

Discussion

Primary hyperoxaluria is a rare autosomal recessive disorder, caused by a functional defect of the liver specific peroxisomal alanine glyoxylate aminotransferase (AGT). The consequence decreased transamination of glyoxylate to glycine leads to a subsequent increase in its oxidation to oxalate, which is a poorly soluble end product. The disorder is characterised by recurrent calcium oxalate nephrolithiasis and nephrocalcinosis, frequently leading to renal failure and death before the age of 20 years. Although more common in early childhood, the age at onset of symptoms ranges from birth to the 6th decade. The disease is classified into two types, type I (glycolic aciduria) and type II (L glycric aciduria), on the basis of the different patterns of urinary organic acid secretion.

Calcium oxalate concentrations are raised in many patients on dialysis, but it is usually only in those patients with primary hyperoxaluria that deposition occurs in extrarenal tissues, despite the use of crystal inhibitors. Postmortem studies have revealed the extent of extrarenal calcium oxalate deposits in this condition, with involvement of bone marrow, cartilage, and myocardium being most common. Crystal deposits have also been described in the prostate gland, testis, thyroid gland, spleen,
liver, thymus, pituitary gland, adrenal gland, pancreas, and the central and peripheral nervous systems. In most of these cases, the deposits were limited to the vessel wall, with arterial and arteriolar involvement being more common than venous involvement.

Reported cases of ischaemic damage secondary to vessel wall involvement include the sudden onset of peripheral ischaemia with livedo reticularis, necessitating amputation of a metatarsal, in a 38 year old man and cutaneous gangrene. This is the first reported case of intestinal infarction secondary to systemic oxalosis. Other possible causes of infarction include mesenteric thrombosis, atheroembolism, and vasculitis. An unusual cause of intestinal infarction in patients with renal failure is necrosis associated with Kayexalate in sorbitol, administered orally in the treatment of hyperkalaemia. In this case, there was good flow within the main mesenteric arteries at the time of laparotomy, and histology of the resected specimen excluded other vascular lesions. Therefore, we conclude that infarction was a direct consequence of microvascular occlusion associated with massive oxalate crystal deposition.

The presumed mechanism of the ischaemia is endothelial damage with subsequent spasm after in situ crystallisation as a result of saturating blood concentrations of calcium and oxalate. Contact of plasma with the crystal surface leads to activation of serum complement, with subsequent triggering of neutrophil mediated endothelial cell damage as a result of the release of toxic oxygen radicals. Platelets are thought to augment the endothelial damage by release of their products, with serotonin being particularly implicated. In our patient, it is possible that her calcium oxalate concentrations in the past had rarely exceeded those required for precipitation, and that it was only the metabolic stresses of a surgical procedure that led to her rapid renal insufficiency. The various treatment strategies are discussed comprehensively elsewhere, but the main aim is early diagnosis and treatment with crystal inhibitors and diuresis to prevent further renal damage. Combined liver–kidney transplantation has produced good results in several patients, with some data suggesting a decline in arterial calcium oxalate deposition post-transplantation. Unfortunately, in our patient the development of systemic complications precluded any assessment for transplantation.

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