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Letters to the Editor

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Nephrogenic systemic fibrosis (NSF): the role of tamoxifen

Dear Sir,
Nephrogenic systemic fibrosis (NSF) is exclusively found in patients with marked renal impairment who have been exposed to gadolinium (Gd) through its use as a contrast agent in magnetic resonance imaging (MRI) studies [1]. The main clinical feature of NSF is thickening and hardening of the skin, and joint contractures are common. There is no confirmed effective treatment for NSF and no current evidence in the literature that spontaneous improvement occurs. Improvement in renal function, either by recovery after an acute kidney injury or through transplantation, is thought to be the most beneficial therapy [1–3], although relapses following re-exposure have been described [3]. We describe the case of a patient who we have successfully managed with tamoxifen.

A 57-year-old male developed end-stage chronic renal failure (ESCRF) secondary to biopsy-proven IgA nephropathy and was commenced on haemodialysis. He was diagnosed with an epidural abscess and required surgical decompression, with good results. A cerebellar abscess was later noted, thought to have originated from the primary site. This was treated conservatively and monitored with regular MRI scans to assess progress. Overall, the patient underwent eight separate MRI examinations in a 12-month period, accumulating a total dose of 105 mL gadoteridol (ProHance®). There are no records detailing the timing of post-MRI haemodialysis, but at that time, it was not a routine practice to reschedule haemodialysis sessions to minimize Gd exposure.

Around a year later, he complained of progressive stiffness of his hands and feet. Examination revealed symmetrical thickening of the skin in his distal extremities with marked limitation of joint movement. NSF was confirmed on skin biopsy.

Physiotherapy was commenced but proved ineffectual, and he refused any further input from services, including any formal assessment from the occupational therapy team. He required admission to hospital due to pain, severe enough to require opiate analgesia, and markedly deteriorating mobility resulting in his being virtually immobile. While an inpatient, he was commenced on oral steroids and tamoxifen, which lead to a dramatic improvement of his condition. He left hospital opiate-free and mobile using only a walking stick, and has successfully coped at home since then.

To date, there are no validated treatments for NSF. The natural history of NSF is of progressive fibrosis, leading to contractures and immobility. In 5% of patients, the disease has an aggressive course that results in death—either directly due to respiratory compromise from pleural or diaphragmatic involvement, or by patients electing to discontinue dialysis due to deteriorating quality of life [4,5]. We suggest that this observation should prompt the need for further, randomized controlled trials into the potential benefit of tamoxifen in this situation.

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Intussusception of the small bowel in an adult associated with nephrotic syndrome

Sir,
Here, we report our observations of a 39-year-old female who presented with progressive lower limb oedema over a 2-week period. She had no significant medical or family history. Physical examination revealed a right-sided pleural effusion and bilateral lower limb swelling. Laboratory investigations confirmed the clinical suspicion of nephrotic syndrome: albumin 14 g/L, creatinine 60 μmol/L, cholesterol 8.8 mmol/L, urinary protein–creatinine ratio (PCR) 997 mg/mmol. Complements were normal, and autoimmune serology was negative.

Percutaneous renal biopsy was performed and demonstrated features consistent with minimal change disease.

She was commenced on fluid and salt restriction and high-dose loop diuretics. She also received prednisolone 1 mg/kg orally [1]. Five days post-renal biopsy, she developed acute left-sided colicky abdominal pain. On

physical examination, her abdomen was tender in the left iliac fossa with no signs of peritonism. The abdominal film was unremarkable. Abdominal ultrasound showed an ileocolic intussusception (Figure 1).

She remained clinically and biochemically nephrotic at this time. She entered the remission phase of nephrotic syndrome between Days 7 and 10 after the initiation of therapy which coincided with the complete resolution of her abdominal pain.

Gastrointestinal disturbances are frequently encountered in the course of nephrotic syndrome. The differential diagnosis considered included renal vein thrombosis, peptic ulcer disease and subacute bowel obstruction.

Fortuitously, at the time of ultrasonography, the patient developed an episode of colicky abdominal pain, and the intussusception could be demonstrated.

Ultrasonography is the diagnostic tool of choice to detect intussusception, although it can be operator dependent or limited by body habitus.

Intussusception causes 'telescoping' of the bowel due to a lead point in the bowel, which in this case is due to incoordinate gut motility and bowel wall oedema.

Intussusception is not infrequently described in the paediatric literature, but the usual cause in adults is secondary to a bowel tumour, which acts as a lead point for the invagination of the bowel [2]. Treatment of the underlying nephrotic syndrome resulted in resolution of the intussusception without the need for any intervention [3,4]. Infusions of albumin have also been described [5].

We conclude that nephrologists should consider intussusception in the differential diagnosis of abdominal pain in the setting of nephrotic syndrome as early recognition may improve prognosis.

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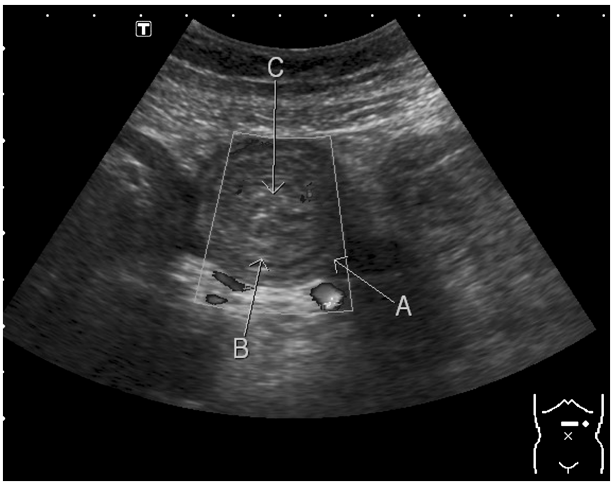

Fig. 1. Transverse section of intussuception. Bull's-eye sign/target/crescent-in-doughnut. **A**. Intussicepiens. Concentric rings of alternating hypoechoic and hyperechoic layers. **B**. Returning limb of intussuceptum. **C**. Mesentery of intussuceptum. Central hyperechoic portion.

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Obstructive uropathy due to inflammatory abdominal aortic aneurysm occurring 18 years after surgical repair of an atherosclerotic aneurysm

Sir,

Chronic periaortitis has a wide range of causes [1]. Obstructive uropathy associated with chronic periaortitis complicating abdominal aortic aneurysm (AAA) repair is well recognized and tends to occur early within 12 months of AAA repair. It is unusual for this to occur many years after surgery.

A 71-year-old male was referred to the renal team after an emergency hospital admission by the medical team with acute kidney injury. Serum creatinine was 774 μmol/L (eGFR 7 mL/min/L), having been 140 μmol/L (eGFR 47 mL/min/L) 4 months previously.

There was history of an atherosclerotic AAA open repair with an aortic graft in 1990. In 1994, the patient had a surgical exploration and evacuation for retroperitoneal haematoma.

In 1995, the patient underwent surgical repair for leaking AAA. The proximal half of the graft was resected and replaced with a second Gelsoft graft (rifampicin soaked).

In September 2008, the patient was admitted to our hospital with malaise, anorexia and oliguria.

On examination, there was a pulsatile abdominal mass just lateral (left) to his midline laparotomy scar. An abdominal CT scan showed a large infrarenal AAA measuring 9.2 cm. The right kidney was of normal size but severely hydronephrotic with a hydroureter extending down to the level of the aortic bifurcation. The left kidney was small and also hydronephrotic (Figure 1). There was extensive fibrosis noted at the level of the aortic bifurcation associated with obstructive uropathy (Figure 2).

The patient remained hypotensive with a poor urine output. He subsequently deteriorated and died 4 days after admission.