Late presentation of ischaemic monomelic neuropathy after vascular access surgery

Introduction

Haemodialysis is an established renal replacement therapy for end-stage chronic kidney disease. In 2014, a total of 24 166 patients in the UK were receiving haemodialysis (MacNeill et al, 2016). Vascular access is essential and while intravenous catheters may be used in the short to medium term, more definitive access usually requires the surgical creation of an arteriovenous fistula or graft. Ischaemic monomelic neuropathy is a rare but devastating complication described in association with surgical vascular access procedures. Ischaemic monomelic neuropathy develops rapidly in the postoperative inpatient period and early recognition is vital to allow potential treatment, although in practice the diagnosis is often delayed (Kirksey, 2010). It is important for those involved in the care of haemodialysis patients to be vigilant for this devastating complication.

Discussion

Ischaemic monomelic neuropathy is a serious neurological complication of vascular access surgery. Precise data on incidence are lacking, although it is rare with limited reporting in the literature. It is most often seen in the upper limbs and is more common with prosthetic grafts than autologous arteriovenous fistulas (Gibbons, 2015). Ischaemic monomelic neuropathy typically affects the distal sensory and motor branches of the ulnar, median and radial nerves resulting in severe hand pain with a distal-to-proximal gradient of multiple neurological deficits (Wodicka and Isaacs, 2010).

Classically, ischaemic monomelic neuropathy presents immediately after vascular access surgery with pain and severe loss of neurological function which appears disproportionate to ischaemic changes affecting other upper limb tissues, such as skin or muscle (Wodicka and Isaacs, 2010). This could be explained by the postulated pathophysiology in which acute transient circulatory occlusion of a limb results in ischaemia which is severe enough to injure nerve fibres but insufficient to damage other tissues (Wodicka and Isaacs, 2010; Tan et al, 2014).

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CASE REPORT

A 58-year-old South Asian man underwent a right upper limb vascular access graft procedure for haemodialysis, despite multiple previous attempts at vascular access including four previously failed arteriovenous fistulae and several clotted femoral and jugular venous catheters. His past medical history included type 2 diabetes mellitus, diabetic retinopathy, ischaemic heart disease and stage 5 chronic kidney disease.

Under general anaesthesia a 6 mm PTFE (polytetrafluoroethylene) graft was anastomosed to the brachial artery just above the elbow crease, tunneled through the subcutaneous tissues of the delto-pectoral region and drained via a HeRO outflow (Merit Medical) component via the axillary vein. Good flows were noted in the graft at end of surgery. While in recovery the patient had a cardiac arrest which responded to immediate cardiopulmonary resuscitation resulting in the return of spontaneous circulation after 4 minutes. He was transferred to intensive care for post-arrest sedation and ventilation. He made an excellent recovery over 48 hours but unfortunately the newly created right arm graft had clotted during his circulatory downtime. Upon full return of consciousness, the patient did not complain of right arm symptoms. However, physical examination revealed weakness and loss of sensation despite the presence of radial and ulnar pulses and good hand perfusion clinically. As the graft had thrombosis, a surgical exploration and ligation was not indicated.

A referral to the neurological clinic was made for further evaluation. Two months after his surgery, distal trophic changes with skin flaking and a dusky appearance of the fingers of his right hand were noted (*Figures 1a* and *b*), with mild wasting affecting right intrinsic hand muscles. There was distal weakness with Medical Research Council (MRC) grade of 1/5 for abduction of the right thumb and fingers, and grade 2/5 for flexion and extension of the right fingers and wrist. Right elbow flexion/extension and shoulder abduction had normal power. The right supinator reflex was absent but the biceps and triceps reflexes were preserved. There was impaired pinprick sensation below the right elbow and vibratory and proprioceptive sensation was lost distally in the right upper limb. Examination of the left upper limb was normal.

Electrophysiological testing of the upper limbs was requested (*Table 1*). Nerve conduction studies revealed absent distal sensory and motor responses from the right ulnar, median and radial nerves. This was suggestive of severe sensory and motor axonal degeneration affecting these nerves in a distal distribution. Needle electromyography (*Table 1*) revealed active denervation in the distal muscles supplied by the right ulnar, median and radial nerves with no evidence of abnormality in the proximal muscles supplied by any of these nerves. This was also consistent with severe axonal degeneration exclusively affecting the distal segments of these nerves in the right upper limb.

A late diagnosis of ischaemic monomelic neuropathy was established with the characteristic distal-to-proximal gradient of monomelic (i.e. single limb) neurological deficits affecting multiple nerves confirmed with electrophysiological studies. Unfortunately, no surgical intervention could be offered as the nerve damage was deemed irreversible.

Figure 1. **a.** Right hand (at rest) compared to the left hand. Note the dusky appearance of the right fingers and skin scaling. **b.** Maximal effort in extending the fingers of the right hand (MRC grade 2).

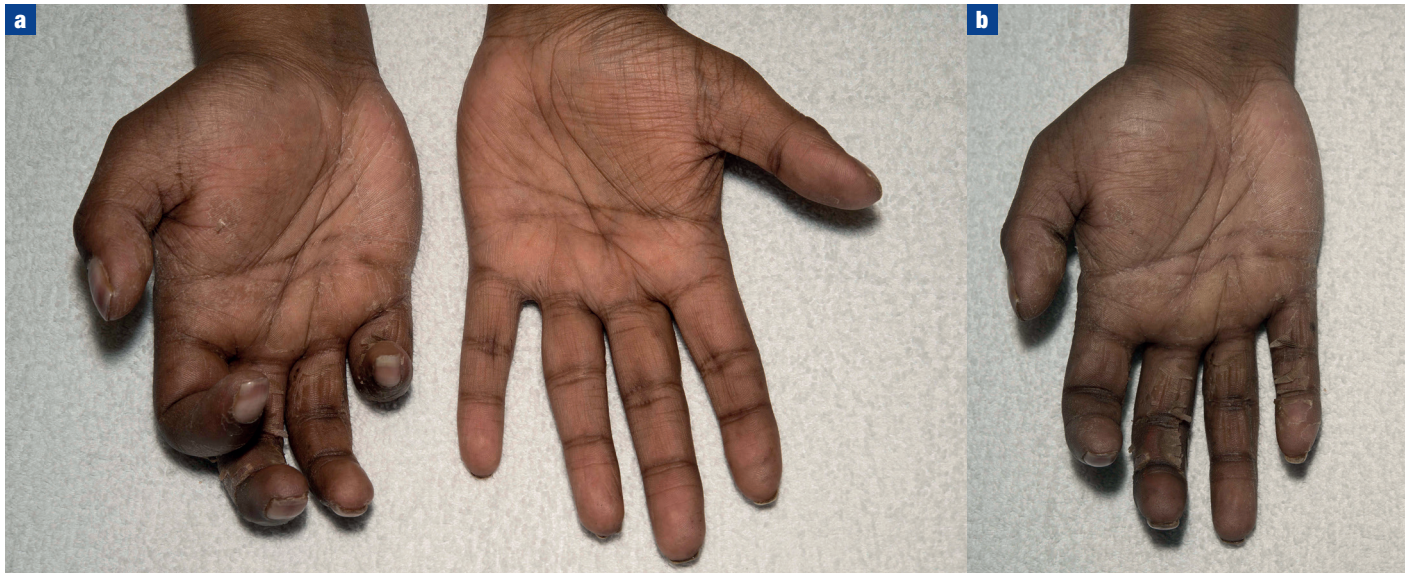

Table 1. Results of nerve conduction studies and electromyography

Nerve conduction studies		Right upper limb	Left upper limb
Motor (normal values)	Median (>4 mV)	Absent	12.5
	Ulnar (>6 mV)	Absent	7.2
Sensory (normal values)	Median (>5 µV)	Absent	9.7
	Ulnar (> 5 µV)	Absent	1.6
	Radial (>12 µV)	Absent	19.2

Electromyography of the right upper limb reveals active denervation in the following distal muscles: abductor pollicis brevis (median), first dorsal interosseous (ulnar), extensor indicis proprius (radial). Electromyography of the following proximal muscles of the right upper limb was normal: flexor digitorum superficialis (median), flexor digitorum profundus (ulnar), triceps (radial), biceps (musculocutaneous) and deltoid (axillary).

An important differential diagnosis of hand pain following vascular access surgery is distal hypoperfusion as a result of arteriovenous-associated ischaemic steal syndrome (Tan et al, 2014). Sensorimotor deficits may be present in arteriovenous-associated ischaemic steal syndrome, but signs of ischaemia in other tissues including pallor, skin ulceration, digital infarction and muscle necrosis will also be present (Miles, 1999; Zamani et al, 2009). Conversely, in ischaemic monomelic neuropathy the hand is warm and pulses are often palpable (Tan et al, 2014).

Prompt recognition and urgent treatment of ischaemic monomelic neuropathy is essential. Immediate ligation of the access is indicated to relieve any residual ischaemia (Gibbons, 2015), although this may not result in full resolution and neurological deficits may be permanent even after urgent vascular intervention (Wodicka and Isaacs, 2010). Clinicians caring for haemodialysis patients should be especially vigilant in initiating rapid treatment for this rare and devastating complication of vascular access surgery (Kirksey, 2010). **BJHM**

LEARNING POINTS

- Ischaemic monomelic neuropathy refers to multiple neurological deficits in a single limb resulting from acute nerve ischaemia, most commonly after creation of an arteriovenous fistula or graft.
- Prompt recognition is vital. Ischaemic monomelic neuropathy should be considered in any patient immediately after vascular access surgery with hand pain, a distal-to-proximal gradient of sensorimotor deficits affecting multiple nerves in one limb, and a paucity of ischaemic signs in other tissues.
- The diagnosis is clinical and waiting for supportive investigations such as nerve conduction studies should not delay management in the acute setting.
- Treatment consists of urgent tie off of the vascular access, although this may not result in resolution. Delayed diagnosis may reduce the chance of reversibility, leading to major disability and reduced function.

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