Living with skin complications from complex regional pain syndrome

KEY WORDS
- Amputation
- Complex regional pain syndrome
- Skin breakdown
- Ulceration

Complex regional pain syndrome (CRPS) is a poorly understood neuropathic condition that can lead to skin breakdown, ulceration and oedema. It is easy to misdiagnose and challenging to treat, as CRPS also impedes healing. Chronic therapy-resistant CRPS can cause irreversible skin breakdown and/or ulceration, resulting in amputation. Here, Victoria Abbott-Fleming, CRPS patients and founder of the charity Burning Nights CRPS Support, shares her story to help raise awareness of this little-known condition.

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Complex regional pain syndrome (CRPS), which was formerly known as reflex sympathetic dystrophy, is a poorly understood neuropathic condition that can affect adults and children. It is important that professionals recognise the signs and symptoms and are aware that CRPS can cause ulcers and skin breakdown. The condition is easy to misdiagnose, especially where skin complications are involved; as a result of misdiagnosis, patients can be accused of self-harming. CRPS is a challenge to treat but patients have a better prognosis when the condition is diagnosed early and treatment started immediately (Carr et al, 2016). It is important to keep limbs moving to improve the blood flow and lessen any circulatory symptoms. It is crucial that a multidisciplinary team is organised to manage patients with CRPS due to the vast diversity of signs and symptoms (Goebel, 2011).

I was first diagnosed with CRPS in 2003. At that time there was very little information available about the condition online or in the medical literature. As a result, I founded the UK charity Burning Nights CRPS Support, of which I am chair. The charity raises awareness of and supports anyone affected by this debilitating condition. Here, I recount my story to give readers a better understanding of the symptoms and how the condition impacts individuals, and also highlight the misunderstandings that can occur.

THE ACCIDENT

In 2002, aged just 23, I qualified as a barrister and was called to the Bar. I knew it was a tough profession, especially for women, but it was what I wanted. To gain experience I decided to lecture in law. Soon after starting in November 2003 I had an accident at work where I slipped down approximately 30 stairs and suffered major soft tissue damage to my right leg, see *Figure 1*. This accident was to change my life forever.

THE MULTITUDE OF SYMPTOMS

I had never really been ill before but the pain I felt from the moment that I stood up after slipping down the stairs hit me like a train. It was like 1,000 knives being pushed into my skin and bone 24/7. I not only had extreme burning then freezing pain in my right leg, but also extreme sensitivity (allodynia and hyperalgesia) to even the most minor changes in temperature, touch or environment. I suffered hair growth and then loss, as well as brittle and ridged nails. The skin on my leg changed colour between red, white and purple, and the temperature of the limb changed wildly from hot to cold. My injured right leg started to sweat excessively and swelled to three times the size of the left. I began to have tremors. The stiffness of my ankle made it very hard to walk properly.

Due to the lack of sleep, constant pain and inability to work, I fell into a deep depression. I did not want to know anyone or do anything. I even

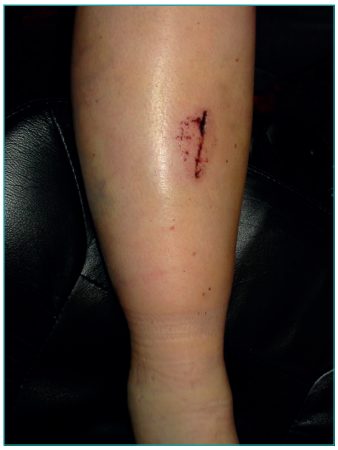

Figure 1. The fall caused major soft tissue damage to the right leg

Box 1. Changes that occur in complex regional pain syndrome (Ott and Maihöfner, 2018)

- Sensory
- Trophic
- Vasomotor
- Autonomic
- Motor
- Disproportionate pain lasting longer than the healing time for the injury.

thought my fiancé, who I met and fell in love with at university, would not want me anymore. I need not have worried about my fiancé, however, as he reassured me that he loved me and would always look after me. He has been my pillar of support throughout the course of my CRPS, for which I cannot thank him enough.

THE DIAGNOSIS

I saw numerous doctors including some who told me it was 'in my head' but I knew that it was not. After 7 months, a pain specialist did some simple tests with a tissue and a blunt hypodermic needle that made me scream in agony. He told me I had all the signs of CRPS (*Box 1)* (Ott and Maihöfner, 2018). CRPS is an extremely painful condition that reaches 42 out of 50 on the McGill Pain Index (Lee et al, 2014). There is no clinical test to diagnose CRPS. Instead, doctors use the Budapest criteria, which use various signs and symptoms from a patient's history and physical examination (Dutton and Littlejohn, 2015).

I had never heard of CRPS and had copious questions. That night I scoured the internet for answers, but there were none. As time passed my main symptoms remained static, but my leg became freezing cold and also changed in colour from red through pink and white to blue, purple and black. This was considered a normal progression of CRPS (Bruehl et al, 2016). I tried the majority of treatments that were available, including medication, physiotherapy, transcutaneous electrical nerve stimulation (TENS), mirror therapy, regional and lumbar blocks, short-term epidural and spinal cord stimulation. All were unsuccessful.

THE START OF SKIN BREAKDOWN

Two years after my accident, the skin on my right leg started cracking and ulceration began. I saw a fantastic tissue viability nurse (TVN). Initially I had Doppler scans showing that I had good, unimpeded blood flow. When I could tolerate it, the TVN tried applying silver dressings, Aquacel, Profore and other such dressings. The problem came when she had to put the dressings on and take them off because I would pass out with the pain. With my allodynia and hyperalgesia, the TVN found it too difficult to treat my leg and to bind the ulcers using the usual compression bandages (Goris, 2015). I felt terrible for her, as she tried so hard to find solutions but every time my leg was dressed, the ulcers would worsen. Just a month after the ulcers first appeared, they had linked to form one big wound. The leg looked angry and was grossly swollen.

Debridement was tried under general anaesthetic (van der Laan et al, 1998) on a number of occasions but the slough would come back almost immediately. The ulcers were so painful that controlling my pain proved almost impossible. I also had recurring infections, which is another complication of ulceration (van der Laan et al, 1998).

My lower leg was now covered in thick, green slough that would constantly drip, see *Figure 2.* It was so bad that I had to use incontinence sheets at home to catch the exudate. I wore toe-post sandals with nappies in them for the exudate and soft skirts because of the slough, allodynia and hyperalgesia. My resourceful mother 'redesigned' my underwear so I would not get any slough on it as I put it on.

At this point, the doctors decided a transfemoral amputation was the only remaining option due to the irreversible skin breakdown, ulceration and infections (van der Laan et al, 1998; Bodde et al, 2011). My above-knee amputation was scheduled for 13 September 2006, when I was aged just 27. It took me a long time to accept my condition and amputation, but when I did I felt that I had really achieved something.

MY LOWEST POINT

During summer 2006 came my lowest point. I got up to find my leg itchier than normal and thought it was moving. As I looked closer I realised there were small maggots all over the sloughy part. I screamed and tears streamed down my face as my husband got me into the bathroom where he added chlorhexidine to buckets of warm water and poured them over my leg. The water felt like acid and I passed out. When I came around, I felt so depressed I wanted to end my life. My husband kept saying that you only have one chance at life and, with his support, I found the strength to carry on and to live with my condition.

KNOWLEDGE AND MISDIAGNOSES

As CRPS is poorly understood, a large number of health professionals did not recognise CRPS, did

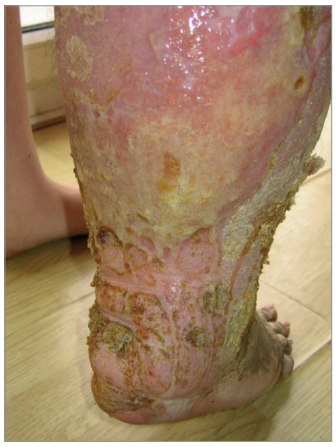

Figure 2. The right leg swelled, ulcerated and the skin broke down irreversibly

Figure 3. Within a week of ulcers appearing in the left leg, the exudate was so bad incontinence sheets were required and an emergency transfemoral amputation was performed

not know how to deal with it, or were unaware that ulceration could be part of the condition. It was extremely frustrating. Following the start of skin breakdown, I was accused of self-harm. Patients with irreversible skin breakdown and/or ulceration, such as myself, are considered to have chronic therapy-resistant CRPS. Many are accused of self-harming (Lipp et al, 1996), however, because of the confusing clinical manifestations. This was not the only symptom that was misdiagnosed. At one stage my oedema was mistaken to be lymphoedema, which I found out was a common misconception.

THE FIRST AMPUTATION

My amputation went ahead as planned, but it took 9 hours and 11 units of blood. Unfortunately the amputation site took 18 months to heal as the CRPS impeded healing. The TVNs and district nurses tried every available dressing, but due to the amount of exudate my husband was changing dressings up to 10 times a day in between nurses' visits.

I was tired and depressed about the pain and the wounds. My husband's support was fantastic, but I felt alone as I had lost many friends because of CRPS and because they did not want to be near someone with wounds.

I had recurring infections once again. In July 2007, the doctors found I had osteomyelitis, so revision surgery was carried out, during which they took another 4 inches off my stump.

SECOND TIME AROUND

After surgery, I kept getting bouts of pneumonia. While on holiday in New York in 2014, I developed swine flu and five types of pneumonia in both my lungs. I did not respond to treatment and after 48 hours my husband was told I had less than a 20% chance of survival. I was given the maximum course of extracoporeal membrane oxygenation (ECMO) and put into an induced coma.

When I finally returned home I began to experience the same pain and other familiar symptoms in my left leg. In March 2014, I was diagnosed with CRPS in my left leg. By November the skin on my left leg had started to break down and ulcerate just like my right leg had done. Within 1 week, the front and side of my leg had completely opened up with ulcers and the familiar smell, exudate and incontinence sheets were back, see *Figure 3*. I could not believe it. On 15 December 2014, 2 days before my 36th birthday, I had an emergency transfemoral amputation.

FINAL WORDS

Although I cannot wear prosthetics, am wheelchair-bound and still have CRPS and phantom limb pain in both stumps, I now have a better quality of life as I do not have the weeping, smelly, ulcerating legs that caused me to be so ill. I know I am extremely lucky to have come through everything. I enjoy my life with my husband and Romanian rescue dog Kipper. And I love what I do now with Burning Nights CRPS Support. WUK

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Burning Nights CRPS Support

The charity provides information about Complex regional pain syndrome (CRPS), along with support for sufferers, carers, friends and families. The charity also has an online forum that acts as a peer support network and a helpline manned by CRPS patients.

Website: www.burningnightscrps.org
Helpline: 01663 795055
10am–4pm, Monday to Friday
Samaritans: 116 123 (outside helpline opening hours)