A case of sympathetically maintained pain

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A 47-year-old woman sustained a gunshot wound to the left thigh. The entry wound was on the anteromedial aspect and the exit wound was on the posterolateral aspect of the thigh. She presented a month later with burning, predominantly superficial pain and cold hyperalgesia affecting part of the left leg and foot in a stocking distribution. She had full pulses bilaterally, numbness to both pin prick and light touch on the sole and lateral aspect of the left foot, wasting of the calf muscles, weakness of dorsiflexion, and cold leg.

Temperature profiles were requested (figures 1 and 2). Magnetic resonance imaging (MRI) scan of the left thigh showed evidence of the previous gunshot wound with the sciatic nerve identified in continuity but deviated at the site of the bullet track and surrounded by scar tissue. The patient had some relief of her symptoms following a left lumbar sympathetic block using a local anaesthetic, but continued to take oral analgesics.

Figure 1 Temperature profile of lower limbs before treatment

Figure 2 Temperature profile of lower limbs after treatment

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Accepted 23 March 1999

Questions

- 1 What abnormality do the temperature profiles show, and what is the condition called?
- 2 What is the definition, and what are the diagnostic criteria for this syndrome?
- 3 What are the former names for these syndromes, and how do they relate to the term 'sympathetically maintained pain'?

Answers

QUESTION 1

The temperature profile (figure 1) shows a drop in temperature affecting the left leg, while (figure 2) shows the improvement in the temperature profile following treatment. The underlying condition is Complex Regional Pain Syndrome, type II (CRPS type II).

QUESTION 2

CRPS type II is defined as burning pain, allodynia (pain from innocuous mechanical or thermal stimuli), and hyperpathia (exaggerated subjective response to painful stimuli with continuing sensation of pain after stimulus has ceased) usually in the hand or foot after partial injury of a nerve or one of its major branches. The diagnostic criteria are:

- the presence of continuing pain, allodynia, or hyperalgesia (lowered pain threshold and enhanced pain perception) after a nerve injury not necessarily limited to the distribution of the injured nerve
- evidence, at some time, of oedema, changes in the skin blood flow, or abnormal sudomotor activity in the region of the pain
- this diagnosis is excluded by the existence of conditions that would otherwise account for the degree pain and dysfunction.

All three criteria must be satisfied.¹

QUESTION 3

The International Association for the Study of Pain, in its second edition on the taxonomy of pain, has replaced the term reflex sympathetic dystrophy with a new term, CRPS type I, while the term causalgia has been replaced by the term CRPS type II.

Sympathetically maintained pain is a type of pain that is maintained by sympathetic efferent innervation or by circulating catecholamines and is a feature of CRPS syndromes and several other types of painful conditions, but it may exist as an entity, not associated with any other condition.²

Discussion

The complications of CRPS are serious and include disuse atrophy of the involved limb, complete disruption of normal daily activity by severe pain, risk of suicide, and drug abuse.³ While a few patients experience spontaneous remission without proper treatment, the majority become progressively worse. Therefore, it is of great importance that the diagnosis of CRPS be made promptly, so that treatment can be started without delay.^{4,5}

The conventional method of treatment for CRPS is sympathetic nerve blockade. This is performed at the level of L₂ for the lower limbs.⁶ Other treatment modalities (listed in no particular order) include percutaneous epidural spinal cord stimulation,⁷ intravenous regional guanethidine,⁸ oral nifedipine,⁹ transcutaneous nerve stimulation,¹⁰ and prostaglandin E₁ ointment.¹¹

Final diagnosis

Complex regional pain syndrome type II.

Keywords: complex regional pain syndrome; pain; causalgia

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Skin fragility and abnormal liver function tests

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A 53-year-old man, a worker in a plastic factory, presented with a 12-month history of a non-itchy skin rash on the dorsum of both hands, associated with increased skin fragility and tendency to recurrent scar formation. The biochemical profile revealed an isolated elevation of serum alanine aminotransferase 55 IU/l (normal 5–36 IU/l). Clinical examination did not reveal any stigmata of chronic liver disease. He denied any history of jaundice, blood transfusions, intravenous drug use or family history of skin or liver disease. His alcohol intake was in the range of three bottles of wine a week. Hepatitis B surface antigen and hepatitis C antibodies (ELISA) were not detected. Serum copper, caeruloplasmin and alpha-1-antitrypsin levels were normal. No serum auto-antibodies were detected. Subsequently, a liver biopsy was performed (figure).

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Accepted 3 December 1998

Figure Liver biopsy (Perls stain, orig $\times 400$)

Questions

- 1 What does the liver biopsy show ?
- 2 What is the underlying skin disorder ?
- 3 How is the condition treated ?