CASE REPORT

The use of regional anaesthetic blockade in a child with recurrent erythromelalgia

C M Harrison, J M Goddard, C D Rittey

Arch Dis Child 2003;88:65–66

Primary erythromelalgia is a rare condition, which is characterised by redness, burning pain, and increased temperature of the extremities. We describe a 6 year old boy with symptoms of erythromelalgia and the difficulty surrounding treatment of this condition. Severe pain responded to the use of regional anaesthetic blocks.

Erythromelalgia is an unusual disease, first described by Mitchell in 1878.¹ Clinically it is characterised by intense burning pain of the extremities, redness, and increased temperature of the limbs.² Initially two types of erythromelalgia were described: a primary type with no associated conditions; and a secondary type, which may occur in many conditions such as myeloproliferative disease, systemic lupus erythematosus, diabetes, and hypertension.^{3,4} Erythromelalgia associated with thrombocythaemia has recently been described as a separate entity.⁵

Primary erythromelalgia occurs more commonly in males, with a male:female ratio of 2:1.³ The literature suggests that one third of patients with primary erythromelalgia are less than 30 years old.² However, there are few previous reports of this condition in childhood. We report a case of primary erythromelalgia in a 6 year old boy who required regional blocks for symptomatic relief.

CASE REPORT

A 6 year old boy, previously well, presented with a three week history of burning pains in his hands and feet, only relieved by immersion in cold water. He had no systemic symptoms and clinical examination revealed erythema of his hands and feet, with peeling of the skin, but with no neurological deficit apart from mild paraesthesia of his feet up to ankle level. Nerve conduction studies (NCS) showed an axonal motor neuropathy with a demyelinating component, characterised by considerably reduced compound muscle action potential and prolonged distal motor latencies.

He was admitted for pain relief and initially treated with simple analgesics, which had little effect. Morphine (10 mg four hourly) and amitriptyline (10 mg three times daily) were therefore added. His symptoms, more in hands than feet, worsened and he remained in hospital. Neurological and pain management teams were involved, and a spectrum of investigations were carried out (table 1).

During the next four weeks, he was given a course of intravenous immunoglobulin (IVIG, 8 g); phenytoin (15 mg/kg loading dose) and carbamazepine (50 mg twice daily, increasing to 150 mg twice daily) were commenced with no benefit. Behavioural therapy was also tried. Intravenous morphine and midazolam infusions were also ineffective. Bilateral axillary brachial plexus catheters were placed, through which bupivacaine (0.25% at 2–4 ml/h) was infused. When pain relief was obtained, topical capsaicin paste was tried. The regional blocks

Table 1 Results of investigations

Investigation	Result
Full blood count	Normal
Erythrocyte sedimentation rate	Normal
Autoantibodies—antinuclear antibody, antimitochondrial	Negative
Rheumatoid factor	Negative
Viral titres, including mycoplasma, borrelia	Normal
Porphyria	Normal
Heavy metals—lead, arsenic, thallium, mercury	Normal
Blood lactate	4.8 (0.9–1.8)
Immunoglobulins	Normal
Oligoclonal bands	Negative
Very long chain fatty acids	Normal
Vitamin B ₁₂	Normal
Folate	Normal
Cerebrospinal fluid glucose	4.1 (2.8–4.4)
Protein	<0.1 (0.1–0.4)
Lactate	2 (0.9–1.8)
Nerve biopsy	Axonal degeneration
Magnetic resonance imaging, brain	Normal

were continued for a week with good pain relief. They were slowly weaned down and he was discharged on haloperidol (0.5 mg twice daily) and capsaicin paste (topically), with regular neurological and psychotherapy reviews. Haloperidol was used because of its recognised benefits in chronic pain management. He appeared to recover and defaulted follow up.

Exactly two years later, now aged 8, the boy re-presented with the same symptoms. His feet were worse than his hands, and symptoms were only relieved by immersion in cold/iced water. Examination revealed red chapped hands and feet with no demonstrable neurological deficit. All the investigations carried out on his first presentation were repeated and were all within normal limits. NCS showed mild motor and sensory abnormalities, identical to, but less severe than the first episode. A short trial of oral analgesics was followed by an epidural catheter and infusion of fentanyl (2 µg/ml) and bupivacaine (0.25%) mixture at 4 ml/h. He improved on this so it was removed after five days, and he was allowed home briefly. He returned with severe pain in his hands and feet, relieved only by hanging them out of the window. His hands and feet were bright red, hot to the touch, and he was unable to weight bear. He was given a further course of IVIG (10 g) and started on gabapentin (300 mg three times daily). He developed weight loss and mild weakness of his interossei. Because of the relief obtained from the cold, he developed cold injury with ulcers on the tips of his fingers and toes. Repeat NCS showed a more severe sensorimotor neuropathy. Many of the

Abbreviations: IVIG, intravenous immunoglobulin; NCS, nerve conduction studies

initial investigations were repeated and remained normal. Bilateral sciatic nerve catheters were placed with an infusion of 0.25% bupivacaine and adrenaline, but fell out after 24 hours. A repeat epidural was performed and attached to an indwelling reservoir system, with an infusion of bupivacaine 0.25% and fentanyl 2 µg/ml. The epidural resulted in complete relief of pain, and it was removed six weeks later. He was discharged home on gabapentin (300 mg three times daily), amitriptyline (25 mg twice daily), and multivitamins.

On review six months after re-presentation, he had decreased sensation in a glove and stocking distribution to all modalities, with mild weakness of the intrinsic muscles of his left hand. Four months later his sensation was improving and he had no residual weakness. At last review, 2½ years after the second presentation, he was off all treatment and had no residual deficit.

DISCUSSION

Erythromelalgia is a rare disease, affecting only three of 5000 patients referred to a pain clinic in one series. Brown⁶ described five diagnostic criteria for erythromelalgia:

- (1) Bilaterally burning pain in extremities
- (2) Sharp increases of local heat in affected parts
- (3) Production and aggravation of distress by heat/dependence
- (4) Relief by cold, rest, and elevation
- (5) Condition refractory to treatment.

Brown also pointed out that the bilateral distribution must be present, although it is not always completely symmetrical.

The pathophysiology of this condition is unknown, but a number of theories have been proposed. These include:

- (1) A lack of sympathetic activity causing lack of vasoconstriction⁷
- (2) Sparseness in autonomic innervation of arterioles and sweat glands⁸
- (3) Decreased concentrations of acetylcholine in nerve terminals of the perivascular plexus in affected people⁸
- (4) Abnormal response to pain modulators⁹
- (5) Neuropathic toxins¹⁰
- (6) Activation of platelets⁹
- (7) Shunting of blood in cutaneous and subcutaneous vessels to subdermal vessels⁹
- (8) Susceptibility of skin to heat¹¹
- (9) Hereditary factors.¹¹

In our patient, the erythromelalgia was associated with a (sensori)motor neuropathy, the severity of which paralleled the severity of the clinical symptomatology. This association has not been previously documented. It is possible that the

mechanism is as described in theories (1), (2), or (5). However, in our case, there was no evidence of neuropathic toxins, and this would favour the first two theories.

As in our patient, the treatment of erythromelalgia is difficult and often ineffective.³⁻⁴ This may be owing to the lack of understanding of the pathophysiology. Behavioural therapies and biofeedback have been attempted but, as in this child, are rarely of benefit themselves. Placebo treatment has been tried in a few studies and no relief has been shown, suggesting that the disorder is not purely psychological. Other treatment modalities attempted (although the results are inconsistent) for symptomatic relief include dry cool air, β blockers, aspirin, sodium nitroprusside, sympathetic lumbar blocks, sympathectomies, and epidural infusions.²⁻³ We avoided those therapies for which data was ambiguous or those with theoretical risk to our patient. In this case, the only effective treatment was regional blockade with local anaesthetic agents, either via bilateral axillary brachial plexus blocks (in the first attack) or lumbar epidural (in the second).

In summary, we present a 6 year old boy who had two separate attacks suggestive of erythromelalgia. This was associated with axonal neuropathy and required axillary and epidural blockade for pain relief

Authors' affiliations

C M Harrison, C D Rittey, Ryegate Children's Centre, Sheffield, UK
J M Goddard, Department of Anaesthesia, Sheffield Children's Hospital, UK

Correspondence to: Dr C Harrison, Jessop Wing Neonatal Intensive Care Unit, Tree Root Walk, Sheffield S10 2SF, UK; c.harrison@doctors.org.uk

Accepted 24 July 2002

REFERENCES

- 1 **Mitchell SW**. On a rare vaso-motor neurosis of extremities, and on maladies with which it may be confounded. *Am J Med Sci* 1878;**76**:17-36.
- 2 **D'Angelo R**, Cohen IT, Brandom BW. Continuous epidural infusion of bupivacaine and fentanyl for erythromelalgia in an adolescent. *Anesth Analg* 1992;**74**:142-4.
- 3 **Levine AM**, Gustafson PR. Erythromelalgia: case report and literature review. *Arch Phys Med Rehabil* 1987;**68**:119-21.
- 4 **Davis M**, O'Fallon W, Rogers R, et al. Natural history of erythromelalgia. *Arch Dermatol* 2000;**136**:330-6.
- 5 **Strozik K**. Case of primary erythromelalgia in a child. *Clin Pediatr* 1992;**31**:378-9.
- 6 **Brown GE**. Erythromelalgia and other disturbances of the extremities accompanied by vasodilatation and burning. *Am J Med Sci* 1932;**183**:468-85.
- 7 **Mufson I**. Clinical observations in erythromelalgia and method for its symptomatic relief. *Am J Heart* 1937;**13**:483.
- 8 **Uno H**, Parker F. Autonomic innervation of the skin in primary erythromelalgia. *Arch Dermatol* 1983;**119**:65-71.
- 9 **Jorgensen HP**, Sondergaard J. Pathogenesis of erythromelalgia. *Arch Dermatol* 1978;**114**:112-14.
- 10 **Martin J-C**. Erythromelalgia: a familial case. Discussion of the role of mercury. *Annales de Dermatologie et Venereologie* 1994;**121**:309-14.
- 11 **Thompson GH**, Hahn G, Rang M. Erythromelalgia. *Clin Orthop* 1979;**144**:249-54.