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Erythromelagia: A Rare and Hard-to-Treat Condition: A 9-Year-Old Boy Responsive to Intravenous Lidocaine and Oral Mexilitene

The authors have no potential conflicts of interest to report.

Dear Editor:

We would like to report our successful use of intravenous lidocaine and oral mexiletine in the treatment of erythromelalgia on a 9-year-old boy.

One month prior to consult, he experienced progressive, burning pain in hands and feet that was relieved with cold water. The affected extremity demonstrated mild erythema, swelling, and warmth. He arrived to our institution derived from another hospital, reporting a pain intensity of 10/10 on the visual analog scale (VAS) with gabapentin as sole treatment, and no diagnosis to explain these signs and symptoms.

Initially, the pediatric unit ordered oral acetaminophen, ketorolac, and pregabaline for neuropathic pain management, but there was no response after 4 days of treatment.

Our pain unit evaluated the patient and performed a bolus injection of intravenous lidocaine (1 mg/kg in 20 mL of 0.9% saline over 1 minute), with good response, decreasing his VAS pain intensity to 3/10. A trial intravenous lidocaine infusion was implemented (4 mg/kg in 250 mL of 0.9% saline over 3 hours). He responded well to 3-hour intravenous lidocaine infusion, and his treatment continued with pregabalin (75 mg every 12 hours), acetaminophen (15 mg/kg every 8 hours), tramadol (25 mg every 12 hours) and amitriptyline (25 mg every 12 hours), intravenous lidocaine (4 mg/kg over 3 hours daily), and intravenous methadone 2 mg as needed.

With this background, a multidisciplinary meeting was held, concluding erythromelalgia as the most likely diagnosis due to clinical symptoms and signs and negative laboratory results.

Because of known reports of erythromelalgia secondary to autoimmune axonopathy, a trial of corticosteroids was started. Intravenous methylprednisolone pulse therapy was administered (30 mg/kg/d) for 3 days and then maintained with oral prednisone 30 mg/day, without response after 4 days of treatment.

Since adults with erythromelalgia who responded to intravenous lidocaine had been successfully treated with oral mexiletine, and there were reports in the literature of the same in children [1] with little or no adverse effects [2], we decided to initiate treatment with mexiletine on our patient.

After 7 days on intravenous lidocaine, an initial oral dose of mexiletine (1 mg/kg) was started. The patient began walking again feeling pain (3/10) only while standing. Five days later he was discharged receiving pregabaline (150 mg every 12 hours), amitriptyline (25 mg in the morning and 50 mg at night), and mexiletine (40 mg every 8 hours).

During his visit as an outpatient, the mexiletine had to be increased twice, first to 60 mg every 8 hours 4 days after discharge, and 2 months later to 80 mg every 8 hours. He was able to carry out normally activities with little pain. From the fourth month onward, he referred pain 0/10 while only on mexiletine as pain therapy. Mexiletine was suspended on the twentieth month of treatment. At his last known visit to the doctor as an outpatient 2 months after suspension of mexiletine, the patient remained asymptomatic.

In erythromelalgia, pain is typically refractory to different therapies, with each patient receiving several drugs without proper relief [1]. A standardized therapy, to which most patients respond, does not exist. There is a wide range of drugs with reports of a good response in patients with erythromelalgia ranging from topical to invasive procedures. Topical drugs, such as lidocaine [3], have worked on some patients. Orally, aspirin is the most recurrent drug used, but its action would be limited to cases secondary to polycythemia and thrombocytosis [4].

There are also reports of the use of anticonvulsants, most notably gabapentin, which had a positive effect in some patients [4]. As a result, it has even been suggested that gabapentin should be considered as one of the first-line drugs in managing patients with erythromelalgia [5]. Our patient, however, did not respond well to it.

Mexiletine has been used successfully for neuropathic pain, particularly in diabetic neuropathy [1]. It has been reported that a good response to intravenous lidocaine predicts a good response to mexiletine [1]. Because erythromelalgia is a rare condition, it still poses many questions, particularly regarding its pathophysiology and its treatment. While there are many reports of good responses to various analgesic methods in the literature, effective therapy for most patients has not yet been established.

Francisca Elgueta, MD,
Juan Carlos de la Cuadra-Fontaine, MD,
Leticia Clede, MD, Claudio Fierro, MD, and
Antonia Valderrama, MD
Pain Management Unit, Division of Anesthesiology, Pontificia Universidad Católica de Chile, Santiago, Chile

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