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CLINICAL INFORMATION

Pain management in children with erythromelalgia: case report

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KEYWORDS
Erythromelalgia;
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Abstract Erythromelalgia is a neuropathic pain syndrome due to an autosomal dominant gene, characterized by erythema, increased skin temperature and burning pain in hands and feet, whose treatment is often unsatisfactory. In this paper, we report a case of a 9 years old female patient whose first episode of burning pain, erythema and edema of the hands, without triggering factors, had instant relief after immersion in cold water. She presented with systemic arterial hypertension and had seizures. The patient was treated with gabapentin (150 mg.8 h^{-1}) and amitriptyline (12.5 mg) orally, intravenous lidocaine infusion (120 mg), without relieving pain complaints. Due to the lack of response to the proposed treatment, it was decided to gradually reduce these medications and to introduce carbamazepine (200 mg) orally and, after 4 days of treatment, there was complete relief of the manifestations.

PALAVRAS-CHAVE
Eritromelalgia;
Dor neuropática;
Crianças

Tratamento da dor em criança com eritromelalgia: relato de caso

Resumo Eritromelalgia é uma síndrome dolorosa neuropática decorrente de gene autossômico dominante, caracterizada por eritema, aumento da temperatura da pele e dor em queimação, em mãos e pés, e o tratamento é muitas vezes insatisfatório. Neste caso, está o relato de uma paciente do sexo feminino, com nove anos e primeiro episódio de dor em queimação, eritema e edema em mãos, sem fatores desencadeantes, com alívio instantâneo após imersão em água fria. Apresentava hipertensão arterial sistêmica e teve crises convulsivas. Foi tratada com gabapentina (150 mg.8 h^{-1}) e amitriptilina (12,5 mg) via oral, lidocaína (120 mg) venosa em

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infusão, sem alívio das queixas álgicas. Devido à ausência de resposta ao tratamento proposto, decidiu-se redução gradativa dessas medicações e introdução de carbamazepina (200 mg) via oral e após quatro dias de tratamento houve alívio completo das manifestações.

Introduction

Erythromelalgia is a chronic and debilitating disease characterized by burning pain and increased skin temperature, associated or not with edema. Usually affects upper and lower extremities, which worsens with heat and improves with low temperatures. It may be primary or idiopathic and is an autosomal dominant inherited disorder. Secondary erythromelalgia occurs in association with vasculopathies, collagen diseases, diabetes mellitus, peripheral neuropathy and use of some drugs,[1] in addition to hypertension, systemic lupus erythematosus, rheumatoid arthritis, HIV, gout, thrombocytopenia, myeloproliferative disorders, occurs mainly in a later phase. Approximately two thirds of cases are primary and one third is secondary.[2]

The clinical manifestations of primary erythromelalgia begin in childhood or adolescence and, with advanced age, it may progress and become constant.[3]

Although not well known, the pathophysiological mechanisms of erythromelalgia include increased blood flow, microvascular shunt, increased local metabolism and small fiber neuropathy. Erythromelalgia is linked to a mutation with an abnormality in ion channel.[4] Histopathological examination reveals reduced density of the autonomic nerve plexuses in the skin.[5]

There is no cure for erythromelalgia; thus, its treatment is focused on relieving the patient's manifestations, which may be done with topical, oral, and behavioral therapy. Erythromelalgia usually follows a chronic course, sometimes progressive and incapacitating.[6]

Erythromelalgia causes impaired quality of life, in addition to higher mortality.

Because it is a rare condition with a severe evolution, its early diagnosis and pain treatment are fundamental. The patients affected may evolve with ischemia and finger and toe gangrene by exposure to cold in an attempt to relieve the pain.

Case report

A nine-year-old girl, 28 kg, presented with a history of severe burning pain in the palmar region for 20 days, which showed instant improvement after immersion in cold water. Hospitalized for five days, the patient and her guardian reported that it was the first occurrence of the symptom.

On physical examination, the patient had good general condition, heart and lung auscultation without alterations, hypertension, present and symmetrical pulses in the upper (radial and ulnar 4+) and lower limbs (bilateral pedis and tibial 4+). In the palmar region, she had bilateral hyperemia, increased local temperature associated with mild edema, without limitation of joint movement angles, strength preserved, without trophic changes.

She denied concomitant diseases or previous hospitalizations and had already sought medical attention at another service seven days ago; prednisone (10 mg.8 h^{-1} for five days) was prescribed orally without symptom relief.

On the day of the evaluation the child received intravenous dipyrone (100 mg every 8 h) without pain relief.

Blood count was unchanged.

Erythromelalgia was diagnosed by the Pain Department team and gabapentin (150 mg.night^{-1}), amitriptyline (12.5 mg.night^{-1}), and dipyrone (750 mg.6 h^{-1}) were prescribed orally. Intravenous lidocaine (120 mg.h^{-1}) was also given once.

The child had three convulsive seizures the day after the medication introduction, with different characteristics. The first one, at 6 h, generalized tonic-clonic, lasted less than two minutes; the second one occurred after one hour, with a fixed stare, not responsive to calls and developed a reduction of oxygen saturation, lasting less than two minutes with spontaneous resolution, followed by drowsiness. The third one occurred after 12 h with horizontal nystagmus and hand tremor, and lasted less than 25 s. Diazepam and phenytoin (intravenous until seizure control) were administered, and the patient was sent to the Intensive Care Unit (ICU) with no need for tracheal intubation.

A magnetic resonance imaging scan of the brain was performed, which showed images suggestive of reversible posterior encephalopathy syndrome, with evidence of areas with cortical and subcortical signal alterations in both brainstems of parieto-occipital predominance, larger on the left, with characteristic distribution favoring the possibility of an erythromelalgia diagnosis.

Gabapentin was maintained and amitriptyline and intravenous lidocaine were discontinued. The child was discharged from the ICU after five days, normotensive and without convulsive seizures, under prazosin (2 mg.12 h^{-1}), propranolol (40 mg.12 h^{-1}), and phenytoin (100 mg.12 h^{-1}).

The gabapentin dose was increased to 150 mg.8 h^{-1} (5 mg.kg^{-1}.dose^{-1}), but the patient remained with severe pain (NPRS 8). After discussing with the neurology team, carbamazepine (200 mg.8 h^{-1}) was introduced, with gradual reduction (one-third of the initial dose every 24 h) of other anticonvulsants (phenytoin and gabapentin), up to discontinuation after 72 h.

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