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Primary erythromelalgia - Case report *

Eritromelalgia primária - Relato de caso

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Abstract: Erythromelalgia is a rare clinical syndrome characterized by heat, redness and intermittent pain in the extremities, being most frequent the bilateral development in the lower extremities. Local cooling brings relief to symptoms, while heating, physical exercises and use of stockings/socks intensify the discomfort. This condition can be primary or idiopathic or secondary to haematological disorders and vascular inflammatory and degenerative diseases. It is reported the case of an eighteen-year-old male who presented, at the early age of two, development of the symptoms of erythema, heat and pain followed by desquamation of hands and feet, in outbreaks, with intervals 4 to 5 years long between the crises.

Keywords: Erythema; Erythromelalgia; Hot temperature; Pain

Resumo: Eritromelalgia é uma síndrome clínica rara, caracterizada por calor, rubor e dor intermitente nas extremidades, sendo frequente o acometimento bilateral das extremidades inferiores. O resfriamento local provoca alívio dos sintomas, enquanto aquecimento, exercícios físicos e uso de luvas e meias intensificam o desconforto. A desordem pode ser primária ou idiopática, ou secundária a distúrbios hematológicos e doenças vasculares inflamatórias e degenerativas. Relata-se o caso de um jovem de dezoito anos, com início precoce aos dois anos de idade dos sintomas de eritema, calor e dor, seguidos de descamação nas mãos e pés, em surtos, com intervalos longos de 4 a 5 anos entre as crises.

Palavras-chave: Dor; Eritema; Eritromelalgia; Temperatura alta

INTRODUCTION

Erythromelalgia, a rare condition described initially by Mitchell in 1878,¹ is characterized by the triad of paroxysmal hyperthermia of the extremities with erythema, pain and intense burning and increase in skin temperature.² The disease manifests itself primarily in the feet and the hands but it can also affect the ears although less frequently.¹ Heat exposure, physical exercises, gravity and the use of stockings/socks and gloves can act as triggers of the symptoms or intensify the discomfort while cold causes discomfort

relief.^{3,5} The disease takes typically a chronic course and it is associated with a decrease in the quality of life and considerable morbidity. It differs from Lane's palmo-plantar erythema or red palms syndrome – a congenital, familial and hereditary dysplasia characterized by persistent and symmetric redness, without subjective manifestations - due to the intermittent nature of the symptoms.⁶ Erythromelalgia can be classified as primary or secondary, depending that if the absence or presence respectively of associated diseases that

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may precede it, coincides with the beginning of the disease or occurs during its evolution.¹ The primary form can be classified in familiar (autosomal dominant) or sporadic, and in juvenile onset (before 20 years of age, frequently before 10 years) or adult onset. Familiar erythromelalgia of juvenile onset is associated with mutations in the gene *SCN9A* that codifies a voltage-dependent sodium channel and it is more frequent in male.^{1,5} Secondary erythromelalgia can be associated with various disorders like thrombocytopenia, polycythemia, myeloproliferative disorders, hypertension, vasculitis, systemic lupus erythematosus, scleroderma, rheumatoid arthritis, Raynaud's disease, HIV and gout.^{1,2,4,5,7,8} It has a late beginning, from the third decade onwards, without preference of sex. The incidence of erythromelalgia estimated in the Norwegian population was from 0,25 to 0,33 per 100.000 inhabitants per year.⁵ As in Olmsted, Minnesota, such incidence was greater, of 1,3 per 100.000 inhabitants per year, being 1,1 and 0,2 per 100.000 inhabitants per year the incidence of the primary and secondary forms respectively.⁷

Etiopathogenesis of erythromelalgia is not known yet but it is admitted the existence of primary or secondary vascular abnormalities that result in endothelial edema, with increase in temperature and blood flow, hypoxia, platelet aggregation and activation, with release of prostaglandins which produces erythema and pain.^{3,5} It was also observed the presence of small-fiber neuropathy.⁹ Its treatment includes drugs that act in the neuropathy such as gabapentin, tricyclic antidepressants and selective inhibitors of serotonin reuptake, and, in vasculopathy, drugs like acetylsalicylic acid, β -blockers and calcium channel antagonists.⁵ However, the responses vary a lot and remission of symptoms is rarely observed.¹

CASE REPORT

Eighteen-year-old male adolescent, Caucasian, single, student, born and coming from Recife-PE sought the dermatology clinic with complaints of episodes of redness, heat and pain in hands and feet, followed by desquamation, with duration of 15 days. The symptoms had started when he was 2 years old, with intervals between the crises varying from 4 to 5 years, when the patient remained asymptomatic. The symptoms worsened with exposition to heat and relieved with local cooling. In two occasions he reported the onset of crises after staying too long in a queue, and also after a football match. He denied systemic symptoms and he did not present any other comorbidities or family history.

Physical examination proved that the patient was in good health only with erythema and heat in palms and soles (Pictures 1, 2 and 3), being requested

laboratory exams and behavioral methods were taught to him. After eight days he returned with strong desquamation in plantar and palmar surfaces, with erythema in the subjacent skin (Pictures 4 and 5), this receded after seven days. Blood count and uric acid were normal. Serology for HIV, ANA and RF was not reactive. The patient had another crisis after 15 days, this time with a less pronounced erythema and resolution of the symptoms in a week.

DISCUSSION

Diagnosis of erythromelalgia is essentially clinical. The disease is characterized by the triad of paroxysmal hyperthermia of the extremities with erythema, pain and intense burning and increase in skin² As for the case presented here, it was reported improvement of the symptoms with local cooling and worsening with heat exposure, facts that corroborated the diagnosis of the disease.

Due to the absence of comorbidities our patient was classified as carrier of the primary form of the disease which is characterized by early onset of symptoms which was, in this case, at the age of two, and greater prevalence in male.

In erythromelalgia, the palmoplantar onset is the most common.⁵ In a retrospective study Davis *et al.* described the location of erythromelalgia in 168 patients, and in 148 patients the feet were affected (88,1%). In 42 patients (25,6%), hands were affected while just one patient had the ears affected.¹⁰

In some patients erythromelalgia is reversible and remissions might last months or years or can even be a complete cure. This confirms the observations of Kalgaard *et al.* that suggest that erythromelalgia is not an isolated disease but that it is a pattern of response of cutaneous microvasculature.⁸ In a prospective study



FIGURE 1: Diffuse erythema on soles



FIGURE 2: Diffuse erythema on plant and dorse of feet



FIGURE 4: Desquamation on palms after eight days from the beginning of the crisis



FIGURE 3: Erythema on palms of the hands



FIGURE 5: Pronounced desquamation on soles after eight days of the crisis

Davis *et al.* observed that erythromelalgia is associated with a neuropathy of small fibers and primary vasculopathies, characterized by an intermittent increase of blood flow, hypoxia, and possibly shunts, with increase in local cellular metabolism.^{9,11} Due to the hypothesis of shunts, substances that alter the distribution of skin blood flow can improve the cutaneous oxygenation and induce symptoms relief. Aspirin, which inhibits platelet aggregation can quickly relieve the secondary symptoms of coagulopathy. Mork *et al.*, in a double-blind controlled placebo prospective

study in 21 patients observed that the analogue of oral PGE1, misoprostol, given on a 0,4 a 0,8 mg/day for 6 weeks, determined a significant reduction of the pain symptoms and local erythema but this decrease did not persist and after three months of treatment no further continuation was required.³

As the responses for the current available treatments varies a lot, it was chosen as a more conservative treatment for our patient who had been presenting long periods between the crises (5 years) and limited duration (two weeks) of the symptoms. □

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