

Primary erythromelalgia - case report

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Sir,

Erythromelalgia is a rare clinical syndrome characterized by a triad of redness, increased temperature, and burning pain primarily in the extremities. The term erythromelalgia, derived from the Greek words for redness and pain in the extremities. It is precipitated by heat, exercise and dependency, and relieved by cold exposure, rest and elevation. The attacks are periodic and can last for various lengths of time ranging from minutes to days.

Erythromelalgia can present as a primary, idiopathic form or secondary to a number of diseases and conditions. Adults are more commonly involved than children and are more likely to have the secondary form. Secondary erythromelalgia is usually associated with myeloproliferative disorders such as essential thrombocythemia and polycythemia vera [1].

Herein, we describe a typical case of a 28-year-old woman with primary erythromelalgia.

A 28-year-old previously healthy woman presented with complaints of episodic redness, heat and pain in feet since two years. Her symptoms were triggered by lying down or warm temperature exposure and were relieved by cooling measures. Extensive diagnostic work-up looking for secondary causes for the symptoms was negative and Blood count and uric acid were normal. Serology for HIV, ANA and RF was not reactive.

The diagnosis of erythromelalgia was made based on details provided in her clinical history supported by maceration in the affected area (Fig. 1). The patient respond to aspirin with good outcome.

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 skin [1,2]. 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 carrying a primitive form of the disease.

In erythromelalgia, the palmoplantar onset is the most common. 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 [3].

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 Friberg et al. that suggest that erythromelalgia is not an isolated disease but that it is a pattern of response of cutaneous microvasculature [4]. Tham and Giles reported 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 [5]. Due to the hypothesis of shunts, substances that alter the distribution of skin blood flow can improve the cutaneous oxygenation and induce symptoms relief [6]. Aspirin, which inhibits platelet aggregation can quickly relief 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

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Figure 1: Maceration on soles.

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 [7].

Erythromelalgia is a rare disease entity that leads to significant functional impairment. Early recognition of this disorder and patient counseling are very important in order to minimize complications. Further research would be helpful to elucidate the underlying pathophysiology and to identify effective therapies.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

REFERENCES

1. Tang Z, Chen Z, Tang B, Jiang H. Primary erythromelalgia: a review. *Orphanet J Rare Dis.* 2015;10:127.
2. Alhadad A, Wollmer P, Svensson A, Eriksson KF. Erythromelalgia: Incidence and clinical experience in a single centre in Sweden. *Vasa.* 2012;41:43–8.
3. Davis MD, Snadroni P, Rooke TW, Low PA. Erythromelalgia: vasculopathy, neuropathy, or both? A prospective study of vascular and neurophysiologic studies in erythromelalgia. *Arch Dermatol.* 2003;139:1337-43.
4. Friberg D, Chen T, Tarr G, van Rij A. Erythromelalgia? A clinical study of people who experience red, hot, painful feet in the community. *Int J Vasc Med.* 2013;2013:864961.
5. Tham SW, Giles M. Current pain management strategies for patients with erythromelalgia: a critical review. *J Pain Res.* 2018;11:1689-98.
6. Liu T, Zhang Y, Lin H, Lv X, Xiao J, Zeng W, et al. A large temperature fluctuation may trigger an epidemic erythromelalgia outbreak in China. *Sci Rep.* 2015;5:9525.
7. Mørk C, Salerud EG, Asker CL, Kvernebo K. The prostaglandin e1 analog misoprostol reduces symptoms and microvascular arteriovenous shunting in erythromelalgia - a double-blind, crossover, placebo-compared study. *J Invest Dermatol.* 2004;122:587-93.

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