Secondary erythromelalgia – case report

Eritromelalgia secundaria: informe de caso

To the editor,

A 56-year-old caucasian female presented to our dermatology department with episodes of erythematoviolaceous dyschromia along the outer side of the left foot. The dermatosis had started 1 year before and the patient was having 2-3 episodes per month, each one lasting from hours to a maximum of 2 days, with variable intervals between the crises. The condition was accompanied by a non-specific local discomfort, although the patient denied manifest pain. Each episode evolved towards total resolution, with no sequelae. At this point, the functional impact caused by the dermatosis was negligible. Previous medical history was unremarkable and there were no new drugs administered. The patient had noticed the complaints were favored by the pending position of the left lower limb and contact with hot water and relieved with ibuprofen and by raising the limb. Physical examination revealed only 2 erythematoviolaceous plaques on the outer side of the left foot, with relatively well defined borders (Fig. 1). An incisional skin biopsy obtained from one of the lesions showed non-specific findings: a capillary proliferation in the dermis, with some ectatic vessels, and a a discrete lymphocytic inflammatory infiltrate. In the following months, thrombocytosis (800 x 10^9/L) and a borderline erythrocytosis (15,4 g/dL) were detected in routine analyses and the patient was referred to hematology consultation. Concomitantly there was a clinical worsening of the dermatosis, with an increase in the frequency and severity of the episodes, including nocturnal, now inducing local pain, warmth and sensation of burning. The dimensions of the left foot plaque increased and the dermatosis progressed, involving both hands (Fig. 2) and feet, this time markedly limiting the functional capacity of the patient. At this point the patient underwent laboratory studies searching for autoimmunity, which were negative. Meanwhile the investigation performed in the context of hematology, with bone marrow examination findings compatible with a myeloproliferative neoplasm, allowed the diagnosis of polycythemia vera with positive JAK-2 muta-


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Erythromelalgia is a rare heterogeneous disorder with a variable spectrum of severity and should be considered in patients with paroxysmal neuropathic pain. It is probably underdiagnosed and its early recognition is crucial to minimize the impact on patients’ quality of life.

References


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