



ERYTHROMELALGIA

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Erythromelalgia is a rare multisystemic disorder characterized by an intense, intermittent burning pain with redness and swelling. This pain is intensified by high ambient temperatures and relieved by cooling the affected area. Furthermore, erythromelalgia is often a dermatologic manifestation of many underlying hematological and connective tissue disorders and its presence warrants further investigation.

While the exact mechanism underlying erythromelalgia is unknown, it is likely related to a genetic mutation coding for a specific voltage-gated sodium channel known as Nav1.7. This alteration causes abnormal responses to stimuli like heat leading to an over-excitable nervous system. Since the body's blood vessels are controlled by the nervous system, some researchers believe this is the cause for the abnormal redness, warmth and burning experienced.

Erythromelalgia is more commonly a dermatologic manifestation of an underlying disease rather than a primary disease process itself. These causative diseases range from systemic lupus erythematosus to platelet disorders to **syphilis**. Furthermore, medications like calcium channel blockers can predispose a susceptible individual to develop erythromelalgia.

An easy way to remember the clinical features of erythromelalgia is to consider this condition to be the opposite of **Raynaud's phenomenon**. Erythromelalgia can be diagnosed clinically based on predetermined criteria and include the following: (a) burning pain in the extremities (b) pain provoked by heat (c) pain alleviate by cooling and elevation of the affected area (d) redness of the affected skin (e) increased temperature of the skin. The most commonly affected region of the body is the extremities, with the legs being involved more often than the arms. Other less common co-manifestations include the appearance of cyanosis in the extremities, livedo reticularis, flushing of the face and areas of necrosis and ulceration.

Finally, the pain pattern is classically described as intermittent and episodic. Some individuals may develop a constant and unremitting pain pattern, which carries a poorer prognosis. Reports indicate that nearly 10% of patients may achieve spontaneous remission of their symptoms, never to be affected again.

The principles of treatment in erythromelalgia are aimed at correcting the underlying cause. It must be stressed that no one-size-fits-all treatment exists for erythromelalgia and when one approach is unsuccessful, another option should be explored. This increases the importance of patience and open communication with ones dermatologist in order to find a therapy that works the best.

Traditionally the mainstay of therapy is composed of oral medications and symptomatic treatment. Aspirin, serotonin reuptake inhibitors and gabapentin have demonstrated the most promise in treating the manifestations of the disease. A few case reports have claimed misoprostol, the prostaglandin analog, and IVIG to be effective. Recently, a study investigating a novel agent, XEN402, that antagonizes the aforementioned Nav1.7 sodium channel may achieve up to 90% reduction in symptoms with a single infusion. Finally, symptomatic treatment with mechanical cooling, ice packs and elevation provides many patients with the support they need to endure a typical episode.

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