Erythromelalgia

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Erythromelalgia is a term that describes the clinical syndrome of red, hot extremities. This rare disorder has been described and characterized in patients seen at the Mayo Clinic in Rochester, Minn, for many decades. The lower extremities are involved more frequently than are the upper extremities, and involvement is often symmetrical. Symptoms can be intermittent or, more rarely, constant. The pain can be severe and debilitating; patients attempt to cool the affected areas. Primary and secondary forms of erythromelalgia have been described. Polycythemia vera and other myeloproliferative disorders may underlie the secondary form. Recent evidence indicates that a small-fiber neuropathy is associated with erythromelalgia. Treatment is difficult; aspirin, nonsteroidal anti-inflammatory agents, anticonvulsants including gabapentin, tricyclic antidepressants, and vasoactive drugs have all been tried. Response varies. Over time, the symptoms can worsen, stay the same, improve, or resolve in approximately equal proportions.