

Coexistence of Raynaud's syndrome and erythromelalgia.

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Letter #1

SIR,-Erythromelalgia is known as the inverse Raynaud's syndrome; the former disease can be induced by treatment of the latter. We report a woman with Raynaud's syndrome and erythromelalgia.

A 40-year-old woman had had, for 5 years, episodes of pallor and blue discoloration of the fingers and toes after cooling and emotional exertion. During these 5 years she also had intolerance to warming of the feet, which manifested attacks of pulsating pain and burning sensations. She had about ten cold and five to seven hot induced paroxysms daily. There were no trophic changes or edema of the fingers and toes. Distal pulsation remained. Adson's and Allen's tests were negative. A cold provocation test induced pallor and blue discoloration of the fingers and toes. A hot provocation test induced pulsating pain and red discoloration of the feet. Biochemical investigation revealed accelerated ADP induced platelet aggregation. The patient was otherwise healthy. She was given nifedipine (40 mg daily), dihydroergocristine (1-5 mg daily), aspirin (750 mg daily), and hyperbaric oxygen sessions (1-7 atmospheres absolute, 45 min, seven sessions). After 3 weeks she was discharged and no longer had either hot or cold induced paroxysms.

I cannot explain with certainty how or why the two conditions started together. Their coexistence may have resulted from the dysfunction of arteriovenous anastomoses which are implicated in both diseases. Pathological expansion of the anastomoses while small arteries and arterioles are in cold spasm leads to an attack in Raynaud's disease, whereas in a warm environment such expansion might cause erythromelalgia.

By G. E. SLUTSKER

Kuibyshev Medical Institute, Post Box 703, Post Office 100, Kuibyshev 443100, USSR

Letter #2

SIR,-Dr Slutsker (April 7, p 853) describes a woman with both Raynaud's syndrome and erythromelalgia and is surprised at the association. However, in a study of 13 patients with Erythromelalgia we have observed an association with another vasomotor syndrome

in 7 cases.[1] In 23 consecutive cases in which the diagnosis was considered, a diagnosis of Erythromelalgia was reached in 13 (9 primary and four secondary, due to essential thrombocythemia in 2 and polycythemia vera in 2). Of the 9 with primary erythromelalgia 4 had primary acrocyanosis and 1 had Raynaud's disease, while among 4 patients with secondary erythromelalgia 1 had Raynaud's phenomenon with digital ischemia and 1 had digital ischemia only. Nailfold capillary microscopy, done in all these patients, revealed only minor and non-specific changes.

The association of erythromelalgia, Raynaud's phenomenon or digital ischemia, and myeloproliferative disorders is not surprising. Myeloproliferative disorders are a cause of secondary Raynaud's phenomenon because of the raised blood viscosity resulting from the increase in cellular elements. Erythromelalgia seems to be common in myeloproliferative syndromes, being the presenting symptom in 26 of 40 cases of thrombocythaemia reported by Michiels et al [2] The explanation is the vasodilator effect of prostaglandins derived from activated platelets. However, in primary erythromelalgia the pathogenesis is unknown and patients with both primary erythromelalgia and Raynaud's disease have small blood vessels that are hypersensitive both to heat and to cold. Possible mechanisms are abnormal liberation of prostaglandins or serotonin. Another possibility is platelet dysfunction, and altered platelet function has been considered in the etiology of both primary erythromelalgia and Raynaud's phenomenon [3,4].

by ISABELLE LAZARETH and PASCAL PRIOLLET
Department of Hypertension,
Vascular Medicine, and Internal Medicine,
H6pital Saint-Joseph,
75015 Paris, France

1. Lazareth I, Fissinger JN, Ptiolltt P. L'etythemtalgie, uo acrosyndrome rare: treize observations. *Presse Med* 1988;17:2235-39.
2. Michiels JJ, Abels J, Steketee J, van Vliet HHDM, Vuzevski VD. Erythromelalgia caused by platelet-mediated arteriolar inflammation and thrombosis in thrombocythemia. *Amt Intern Med* 1985;102:466-71.
3. Jorgensen HP, Sondergaard J. Pathogenesis of erythromelalgia. *Arch Dermato* 1978; 114:112-14.
4. Hutton RA, Mikhalidis DP, Bernstein RM, Jeremy JY, Hughes G, Dandona P. Assessment of platelet function in patients with Raynaud's syndrome. *J Clin Pathol* 1984; 37: 182-87