

Clinical Images

Iloprost Treatment of Finger Gangrene in a Patient with Raynaud's Syndrome

Yair Levy MD, Yaniv Sherer MD and Yehuda Shoenfeld MD

Department of Medicine B, and Center for Autoimmune Diseases, Sheba Medical Center, Tel-Hashomer, and Sackler Faculty of Medicine, Tel Aviv University, Israel

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A 67 year old woman was admitted due to severe pain in the distal phalanx of the middle finger of her right hand that had lasted for 2 weeks. Her history was remarkable for Raynaud's syndrome of one years duration for which she was treated with nitrates and calcium blockers, for cyclic neutropenia, and for bleeding episodes following a tooth extraction and cataract surgery. Physical examination disclosed gangrene of the tip of her finger [Figure A], and laboratory tests showed prolonged partial thromboplastin time (76 sec). Further tests confirmed the presence of hereditary factor XI deficiency (homozygous type II/II) and ruled out acquired factor XI inhibitors. Treatment was promptly started with the prostacyclin analogue iloprost (50 µg/day) for 10 days. This treatment resulted in a marked improvement of the finger gangrene [Figure B].

Iloprost was found effective in the treatment of Raynaud's phenomenon secondary to systemic sclerosis. Continuous iloprost infusion (0.5-2.0 ng/kg/min) for 6 hours on 5 consecutive days resulted in complete healing of all cutaneous lesions 10 weeks after the treatment in six of seven patients, compared with none of four patients receiving placebo [1]. It was also associated with evidence of prolonged physiologic improvement (including frequency, duration and symptoms of Raynaud's syndrome). In another study, iloprost was infused at the same rate for 8-13 days [2]. In these 12 patients with systemic sclerosis the effect of iloprost was cessation of

imminent gangrene followed by healing in 2 patients, complete healing of ischemic ulcers in 4 patients, and partial healing in 2 patients. Two other patients with Raynaud's phenomenon showed an improvement, while there was no improvement in a patient with vasculitis of the lower leg, and one patient discontinued the treatment [2]. Recently, iloprost was found to be beneficial for another medical condition as well - primary pulmonary hypertension. A daily dose of 100-150 µg of aerosolized iloprost given to 24 patients with this diagnosis resulted in a decrease in pulmonary vascular resistance and mean pulmonary arterial pressure, while it increased cardiac output [3].

The case presented here also emphasizes, as we have reported before [4], that an inherited bleeding disorder (i.e., factor XI deficiency) cannot be considered to have a major protective role against thromboembolic phenom-

ena, or as in this case, against ischemia. Literature reports of thromboembolic phenomena in patients with coagulation factors are scarce, e.g., the report of a patient with factor XI deficiency and myocardial infarction who also had

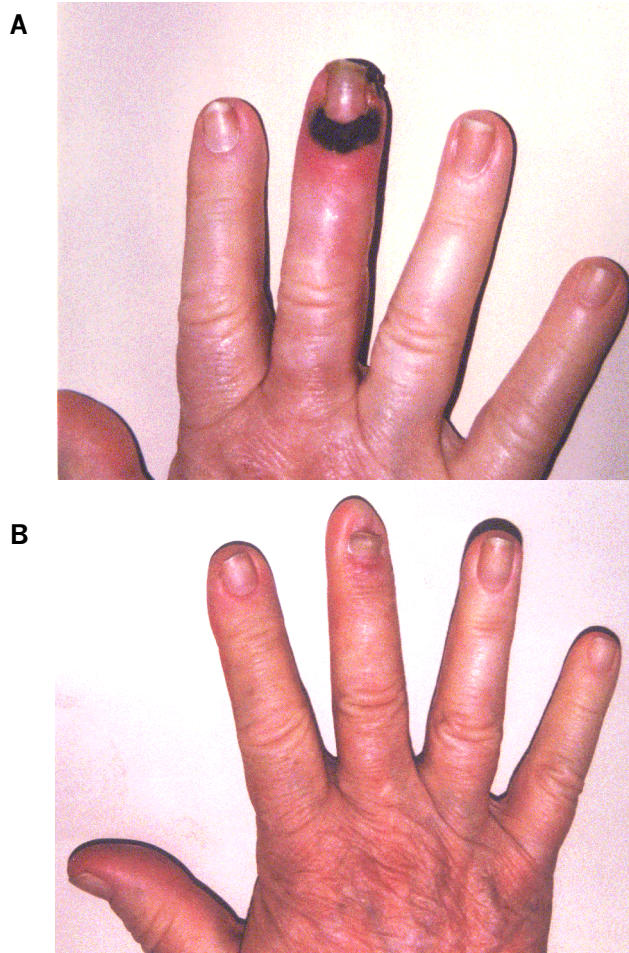


Figure 1. Tip of a finger of a patient with Raynaud's syndrome [A] before, and [B] after administration of iloprost.

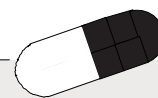
positive lupus anticoagulant test [5]. Therefore, it is possible that on the one hand a patient with factor XI deficiency might have bleeding episodes and also require fresh frozen plasma before an operation, while the same patient could have ischemia due to vascular occlusion or thrombosis on the other.

References

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Correspondence: Dr. Y. Shoenfeld, Dept. of Medicine B, Sheba Medical Center, Tel-Hashomer 52621, Israel. Phone: (972-3) 530-2652, Fax: (972-3) 535-2855, email: Shoenfel@post.tau.ac.il

Capsule



Protein-packed DNA

When rod-shaped bacteria like *Bacillus subtilis* form spores, each developing spore must receive an intact chromosome. Bath et al. looked at how DNA is transferred to spores and found that a protein known as SpoIIIIE appears to act as a sort of DNA pump that actively moves

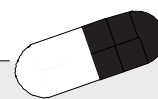
one of the replicated chromosomes into the spore. Since many bacteria possess homologous proteins, such DNA motors may turn out to be ubiquitous.

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Curse the blasted, jelly-boned swines, the slimy, the belly-wriggling invertebrates, the miserable sodding rotters, the flaming sods, the snivelling, dribbling, dithering, palsied, pulseless lot that make up England today.

D.H. Lawrence, English novelist and poet (1885-1930), on a publisher's rejection of Sons and Lovers

Capsule



No immunity to costs

The cost and benefits of activating an immune response are the subject of two reports. Moret and Schmid-Hempel have manipulated a model to measure the costs of innate immune responses in insects. Starved bumblebees stimulated to generate a response to soluble lipopolysaccharide and to sterile particles soon died because they diverted all of their remaining resources to the immune response. In well-fed field populations, most bumblebees carried an infection but showed normal behaviors and activities, which indicates that compensatory mechanisms effectively countered any costs to mounting an immune response.

Promiscuous sexual behavior presents an obvious risk to the individual because of the increased likelihood of contracting a sexually transmitted disease. Although

behaviors may adapt to avoid this hazard, a study by Nunn et al. suggests that in some primate species, it is the immune system that has adapted to cope with the problem. Several species of captive primates were tested for numbers of white blood cells – the mediators of immune defense. The authors found that while there was no correlation of WBC counts with social factors such as group size, frequencies of these cells were significantly elevated in the females of species that had multiple mating partners. The WBC counts of (non-captive) humans were low on this scale.

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