Phenylephrine

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Cutaneous microvascular occlusion syndrome in a patient with a heterozygous factor V Leiden mutation: case report

A 45-year-old man with a heterozygous factor V Leiden mutation developed cutaneous microvascular occlusion syndrome during treatment with phenylephrine.

The man, who had diabetes mellitus and hypertension, was admitted to a neurosurgical ICU following a vertebrectomy. He had undergone numerous neurosurgical procedures and experienced postoperative spinal degeneration and infectious complications; he had also received ventilatory and vasopressor support over a 4-month period and had received norepinephrine [noradrenaline]. However, in the last month, his only vasopressor exposure had been a 2-day course of phenylephrine [at a dosage titrated to maintain target mean arterial pressure; dosage not stated]. One day after phenylephrine cessation, he developed a rash. On evaluation, 4 days later, he had a purple discolouration of his hands and feet. Medications included levofloxacin, quinupristin/ dalfopristin, hydrocortisone succinate, propofol, midazolam, electrolyte supplementation, furosemide, hydroxyzine and metoprolol. He had also received heparin. Physical examination showed symmetrical, well-demarcated purpura and several haemorrhagic bullae on his hands and feet (glove and stocking distribution). He also had retiform purpura on his antecubital fossae and thighs. He had a platelet count of $105 \times 10^3 / \mu L$ and a D-dimer level of 1.9 mg/L. A skin biopsy showed fibrin microthrombi in the superficial and middle dermal vessels with minimal inflammation. He was diagnosed with microvascular occlusion syndrome. The second day after evaluation, his condition started to spontaneously improve. He received lepirudin and his purpura resolved and superficial sloughing yielded mildly erythematous erosions. He did not experience any further thrombotic complications and, 2 weeks later, his condition had nearly completely resolved with no long-term sequelae. Further investigation revealed a heterozygous factor V Leiden mutation. He had an erythrocyte sedimentation rate of 53 mm/h, a C-reactive protein level of 27 mg/L and a functional protein S value (during acute event) of 36% of the normal level. Subsequent functional protein S testing, 2 weeks after resolution, revealed a normal level.

Author comment: "Considering the physiologic effects and temporal relationship of phenylephrine therapy to the onset of our patient's microvascular occlusion, we propose that phenylephrine-mediated vasoconstriction superimposed on his thrombotic predisposition precipitated a transient [microvascular occlusion syndrome]."

Kalajian AH, et al. Phenylephrine-induced microvascular occlusion syndrome in a patient with a heterozygous factor V Leiden mutation. Archives of Dermatology 143: 1314-1317, No. 10, Oct 2007 - USA 801097112