
Cardiovascular Mystery Series

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Pulvis de sclerotium and the mummified finger 1

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October, 1951, and hamlets throughout the south of France were still in the throes of economic recovery; the struggle in Vietnam further drained French resources. Despite postwar scarcities, Fall’s harvest had provided a good yield — an outcome in doubt given Spring’s heavy rainfall and June’s hot sun. Sustenance for destitute Mr. H., his wife and two children, depended on pickled pork, cheese, honey and well water. Spirits soared when they learned baker J. would provide them flour. Though mottled gray and having a sticky texture that made working batter difficult, H. beamed when bread tasted fine. Within days all were brought to clinic, where Nicole Monier, resident in internal medicine, was working. In wife and children, throbbing calves gave way to burning pains and discolored toes. For H., painful fingers turned black. As Nicole examined his right index finger, it fell off! What caused this affliction in previously healthy people? None had fever and others in their village had not taken ill.

This vivid experience remained with Nicole for years. Comparisons were inevitable. For example, Mr. G., a 25-year-old Polish immigrant whose religious preference made him a target of the Nazi reign of terror. His family had fled to the French countryside, where they followed their religious customs and diet. An office clerk, G., complained of a painful right index finger and left thumb worsened by cooler weather. Weeks ago his right hand became swollen and painful. No trauma, fever or chills. Months earlier he had a painful right calf on whose medial aspect was a several centimeter long red cord. The left calf was involved weeks later. A gangrenous digit of the right hand had been lost last winter while stacking firewood. Only in Poland had he smoked cigarettes. Blood pressure normal; heart and abdomen unremarkable. After Nicole had compressed his right radial and ulnar arteries, she instructed him to open and close his hand until it blanched; restoration of color was rapid upon release of the radial artery, but delayed when this maneuver was repeated for the ulnar artery. Dupuytren’s contractures bilaterally. Normal renal function. Occlusions of digital arteries and right ulnar artery were found on arteriography. But why? Vasospasm due to a circulating substance? And then there was Mr. S., a 45-year-old service station worker. For 1 month he noted pain and numbness of 4th and 5th digits of his left hand with blanching, followed by bluish discoloration and then throbbing redness; cooler temperatures were provocative. In changing tires or fixing dented fenders, he used a hammer or his left hand. Normal examination except cool 4th and 5th digits of the left hand with Dupuytren’s contraction. A cord-like mass was felt radial to hypothenar muscles; ulnar pulse was diminished and Allen’s test positive. Arteriography revealed sparse vascularity of digits with aneurysm of the ulnar artery adjacent to the hamate bone. Was a local substance responsible for vasospasm, released perhaps from injured tissue?

Answer

In the Middle Ages, gangrenous ergotism appeared west of the Rhine River, the convulsive form to the east. An outbreak of ergotism appeared in southern France some 40 years ago. With the gangrenous form, limbs and/or digits become swollen; excruciating pains follow. Numbness occurs suddenly and affected parts turn black; mummified, they are lost without pain or hemorrhage. A moist Spring and dry June favor contamination of rye by the fungus Claviceps purpurea whose hard, purple body, or sclerotium, gives infected plant ovaries a cock spur (or ergot) appearance. Midwives gave ergot to hasten child birth; the alkaloid ergotamine was used for migraine headache and pruritus of hepatic origin. Toxic effects, due to intense arterial constriction, resembled those seen in H. and family.

Thromboangiitis obliterans (Buerger’s disease), a vascular disease of lower and/or upper extremities, occurs predominantly in young men whose ethnic origins may

1 A brief mystery which sets the stage for the accompanying mini-review, seeking to integrate basic laboratory and clinical sciences and diverse expressions of disease, while highlighting the role of the generalist (that is, the integrator).
include northern Slavic countries (e.g., Askenazium). Sour rye bread is a dietary staple of these people; smoking is not a consistent habit. Migratory superficial thrombophlebitis is frequent. The hammer hypothenar syndrome involves the superficial palmar branch of the ulnar artery. Often associated with Raynaud’s phenomena, it appears when the hand is used as a hammer. The fibrocontractive disorder of palmar fascia (Dupuytren’s) is based on α-smooth muscle actin containing myofibroblasts whose contractility is induced by peptides (e.g., angiotensin II and endothelin-1). Such substances, if released by these cells, could promote vasospasm of involved vessels.