Aspirin-responsive painful red, black toe, or finger syndrome in polycythemia vera associated with thrombocythemia.

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Abstract Only

Five patients with red, purple blue, or black toes or fingers due to thrombocythemia associated with polycythemia vera (polycythemia and thrombocythemia vera) in four and essential thrombocythemia (thrombocythemia vera) in one are described. The microvascular erythromelalgic syndrome of thrombocythemia was overlooked and progressed to cold blue swollen and painful fingers or black toes in three patients with polycythemia and thrombocythemia vera due to arteriographically documented occlusions of digital or large peripheral arteries with no evidence of preexistent atherosclerotic vascular disease. Concomitant erythromelalgia of the hand palm could be confirmed by the histopathological findings of arteriolar thrombotic lesions in the reticular dermis in two patients with polycythemia and thrombocythemia vera. The increased hematocrit in the presented patients with polycythemia and thrombocythemia vera contributed to the progression of the microvascular syndrome of thrombocythemia to major occlusive ischemic events of the extremities. Standard therapy with oral anticoagulants and reduction of the hematocrit to normal by bloodletting did not affect the platelet-mediated microvascular erythromelalgic, ischemic symptoms in the patients with polycythemia vera because thrombocythemia vera persisted. Complete relief of pain and restoration of the ischemic acral circulation disturbances in patients with thrombocythemia vera or thrombocythemia associated with polycythemia vera in maintained remission by bloodletting could be obtained by long-term treatment with low-dose aspirin.

Keywords Polycythemia vera - Thrombocythemia vera - Erythromelalgia - Arterial thrombophilia - Platelets - Aspirin

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