A Case of Polycythemia Vera with Splinter Hemorrhages

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Once splinter hemorrhage can be considered as a pathognomonic sign of subacute bacterial endocarditis. But it can also be associated with a variety of systemic disorders that increase capillary fragility or primary nail bed involvement in dermatologic disorders. The cause of splinter hemorrhage can usually be established by careful history and physical examination. We report a case of 33-year-old man with splinter hemorrhages, who had polycythemia vera. (Ann Dermatol 14(4) 207~209, 2002).

Key Words : Polycythemia vera, Splinter hemorrhage

Splinter hemorrhages appear as thin black lines, 1-2mm long, located in the distal third of the nail bed. Although the most frequent cause is trauma, it can also be associated with a variety of systemic disorders that increase capillary fragility or primary nail bed involvement in dermatologic disorders[1]. Examples are collagen vascular disease, meningococcemia, eczema, exfoliative dermatitis, psoriasis and cutaneous T-cell lymphoma. The cause of splinter hemorrhage can usually be established by careful history and physical examination. We report a case of 33-year-old man with splinter hemorrhages, who had polycythemia vera.

CASE REPORT

A 33-year-old male presented painful swelling of the right hand with numbness for 1 month(Fig. 1). On physical examination, splinter hemorrhages were noted on the distal third of his nail beds except fifth finger (Fig. 2). Recently he suffered from pruritus after a warm bath or shower, but not treated. He had no episode of trauma on both hands. Routine laboratory tests, autoantibodies and X-ray of both hands were normal except for the elevated white blood cell count of 12700/mm^3, hemoglobin of 19.7g/dL and hematocrit of 63.4%. For differential diagnosis of causing erythrocytosis, further evaluations were done. Laboratory results included normal saturation of O_2, elevated uric acid of 8.4mg/dL, LAP score of 351, Vitamin B12 >2000pg/ml and decreased erythropoietin <1mIU/ml. Abdominal ultrasonography showed splenomegaly measuring above 15cm. Bone marrow biopsy revealed hypercellularity with erythroid hyperplasia and increased number of megakaryocytes. Chromosomal study was normal. Chest X-ray, EKG and echocardiography showed no evidence of subacute bacterial endocarditis. He was in close observation after a phlebotomy under the diagnosis of polycythemia vera.

DISCUSSION

In splinter hemorrhage, blood that is confined to thin grooves between the nail beds longitudinal epidermal ridges adheres to the nail plate and grows out with time. Splinter hemorrhages appear as thin black lines, 1-2mm long, located in the distal third of the nail bed. Although the most frequent cause is trauma, it can also be associated with a variety of systemic disorders that increase capillary fragility or primary nail bed involvement in dermatologic disorders[1]. Examples are collagen vascular

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Fig. 1. Swelling and splinter hemorrhages on fingers of the right hand.

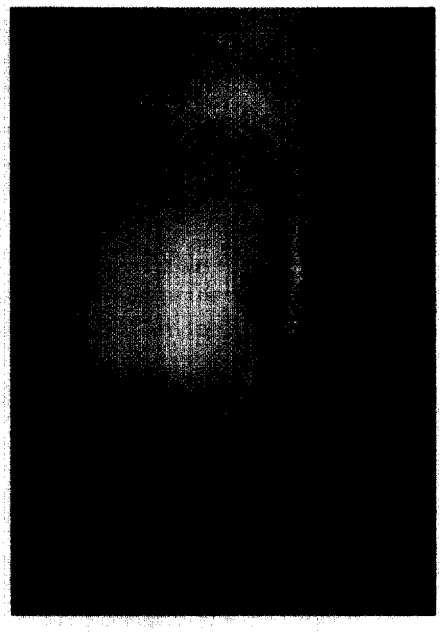

Fig. 2. Splinter hemorrhage on the distal third of nail bed.

disease, meningococcemia, eczema, exfoliative dermatitis, psoriasis and cutaneous T-cell lymphoma. Recently, association with antiphospholipid syndrome[2,3] and vasculocclusive disorder such as thromboangiitis obliterans[4] have often been reported. Rarely splinter hemorrhages may occur as proximal part of the nail bed in trichinosis and vasculitis. Subungal hematoma, more extensive than splinter hemorrhage, may be caused by blood dyscrasias, emboli and scurvy[1].

Polycythemia vera is a myeloproliferative disease characterized by the sustained persistence of an increased level of hemoglobin and an increased hematocrit, not associated with or controlled by erythropoietin. About 30 % have nonspecific symptoms of headache, dizziness, fatigue and parethesias. Other clinical features are splenomegaly, hepatomegaly, hypertension and hyperuricemia[5]. About one-half of patients have peripheral vascular complaints. Manifestations included digital redness or cyanosis, erythromelalgia, digital ischemia with palpable pulses, thrombophlebitis, and coronary or cerebral ischemia[6]. They state that the increased cellular elements led to increased viscosity with subsequent impairment in blood flow, stasis and tissue hypoxia. Aquagenic pruritus develops as well, and rarely livedo reticularis as presenting sign[7].

Our patient was compatible with the diagnostic criteria used by the Polycythemia Vera Study Group[5]. He had all three major criteria: (i) increased red cell mass; (ii) normal arterial O_2 saturation; and (iii) splenomegaly plus two of the minor criteria: (i) leukocytosis; (ii) LAP score >100 or Vitamin B_{12} >900 pg/ml. Also he had aquagenic pruritus and indirect signs of circulatory disturbance of the right hand. It was logical to expect its pathogenesis similar to other signs of disturbed peripheral vasculature.

In conclusion, based on our patient's clinical presentation, it raises the possibility that polycythemia vera may be associated with splinter hemorrhage. Although clinical usefulness is limited, we believe the presence of splinter hemorrhages may lead to an early sign or a diagnostic clue of disease.

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