Cryoglobulinemia with a Digital Gangrene from Monoclonal Gammopathy of Undetermined Significance

Chan Hong Jeon
Division of Rheumatology, Department of Internal Medicine, Soonchunhyang University Bucheon Hospital, Bucheon, Korea

Type I cryoglobulin is composed of a monoclonal immunoglobulin (Ig) and can be found in patients with lymphoproliferative disorders. Some frequent symptoms of type I cryoglobulinemia are skin necrosis and Raynaud’s phenomenon [1]. Monoclonal gammopathy of undetermined significance (MGUS) is a precursor lesion for multiple myeloma [2] and one of the principal causes of cryoglobulinemia [3].

A 37-year-old-male was admitted with gangrene of the left second toe (Figure 1). The patient was diagnosed with primary Raynaud’s syndrome at a tertiary center and has been under observation without treatment for 3 years. Concerning the complete blood count, the white blood cell count was 9,920/μL and the differential was normal. Hemoglobin was 11.5 g/dL; and the platelet count was 361,000/μL. Erythrocyte sedimentation rate was 120 mm/h. Total protein was 10.1 g/dL; and albumin was 3.4 g/dL with a reversed albumin/globulin ratio of 0.5. Serum creatinine was 1.2 mg/dL; and the total calcium was 8.7 mg/dL. The rheumatoid factor was 17 IU/mL; and the antinuclear antibody test was positive but was in a lower titer of 1:40. The test for anti-neutrophil cytoplasmic antibody was negative; and both IgG and IgM anticardiolipin antibodies were negative, and the lupus anticoagulant was also negative. However, cryoglobulin came out as positive. Furthermore, a peripheral blood smear showed marked rouleaux formation and numerous

Figure 1. There was gangrene of the left second toe. Other toes also showed mild cyanotic change on their tips with a background of livedo reticularis.

Figure 2. On peripheral blood smear, after incubation in room temperature, extracellular pale pink amorphous materials were observed (Wright-Giemsa stain, ×200).
Figure 3. A bone marrow biopsy showed a normal myeloid to erythroid ratio and the usual maturation pattern but multifocal aggregates of plasma cells were observed (Wright-Giemsa stain, ×400).

Pale pink amorphous deposits were observed (Figure 2). Because the serologic tests for hepatitis C virus and human immunodeficiency virus were negative and cryoglobulin from paraproteinemia was suspected, protein electrophoresis and immunofixation were performed. The serum M protein was 1.32 g and monoclonal gammopathy of the IgG-lambda type was noted: the ratio of the kappa to lambda free light chain was 0.16. On bone marrow biopsy, myeloid to erythroid ratio was 1.6:1 and showed the usual maturation pattern with multifocal aggregates of plasma cells (Figure 3). The fraction of plasma cells was 8.8%. Chromosomal analysis showed no apparent cytogenetic abnormalities. On skeletal survey, no osteolytic lesion was found.

Based on all the above findings, the patient was diagnosed with cryoglobulinemia from light-chain MGUS [2]. The digital gangrene was treated conservatively with antibiotics and regular dressing. We didn’t plan any additional management for cryoglobulinemia because there was no life-threatening complication, such as hyperviscosity [1]. As for MGUS, since there was no evidence of multiple myeloma or Waldenstrom’s macroglobulinemia, the patient is under periodic follow-up with serum protein electrophoresis and a complete blood count [4].

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES