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Primary systemic amyloidosis with sole cutaneous involvement

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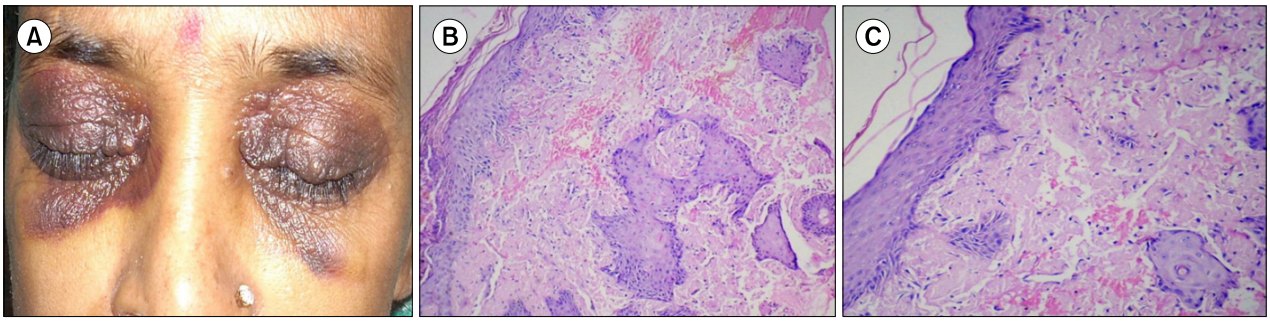

A 60-year-old woman showed blackish pigmentation around both eyes for 2 years, with clinical examination revealing waxy hyperpigmentation and small nodules **(A)**. Blood examination revealed Hb, 7.5 g/dL; WBC, 6.2×10^9/L; platelets, 210×10^9/L; corrected reticulocytes, 1.4%; plasma fasting glucose, 81 mg/dL; serum urea, 26 mg/dL; creatinine, 0.9 mg/dL; total protein, 8.92 g/dL; albumin, 3.96 g/dL; globulin, 4.96 g/dL; albumin/globulin ratio, 0.8:1; calcium, 12.7 mg/dL; and 24-hour urinary protein, 75 mg. A peripheral blood smear showed normocytic, normochromic RBCs with rouleaux formation. Serum protein electrophoresis showed a monoclonal band in the gamma globulin region; serum immunofixation electrophoresis showed monoclonal lambda light chain immunoglobulin. Serum kappa and lambda light chain levels were 6.34 and 626.24 mg/L, respectively, with an altered kappa/lambda ratio (0.01; reference: 0.26–1.65). Radiographic results were negative. Abdominal ultrasonography revealed normal renal corticomedullary differentiation; echocardiography findings were normal. Nerve conduction velocity was normal in all limbs. Punch biopsies from periocular lesions showed amorphous pale pink dermal deposits (hematoxylin-eosin staining; **B**, lower magnification; **C**, higher magnification); Congo red staining showed apple-green birefringence on polarizing microscopy. The bone marrow plasma cell proportion was 49%. Conventional cytogenetics indicated a normal karyotype. Thus, primary systemic amyloidosis may present as isolated skin lesions.