A 66-year-old man presented with a two-year history of fatigue, paresthesia of the legs and feet, weight loss, and shoulder enlargement, with limitation of movement. Examination showed periorbital ecchymoses. What is the most likely diagnosis?
A 66-YEAR-OLD MAN PRESENTED WITH A TWO-YEAR HISTORY OF FATIGUE, paresthesia of the legs and feet, weight loss, and shoulder enlargement, with limitation of movement. On physical examination, periorbital ecchymoses (the “raccoon” sign) and infiltration of the periarticular tissues of the shoulders were found. A biopsy specimen of abdominal fat that was stained with Congo red was positive for amyloid, and serum monoclonal paraprotein (lambda light chain) was detected by immunoelectrophoresis. A bone marrow biopsy specimen contained 30 percent plasma cells. The patient was enrolled in a chemotherapy protocol but died two months later. Although amyloid infiltration around articular structures is rare, the “shoulder pad” sign that results from amyloid deposition in periarticular soft tissue is pathognomonic for immunoglobulin amyloidosis. It has been suggested that kappa III variable light-chain amyloid proteins have an increased predilection for soft-tissue deposition.

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