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Raccoon eyes sign: unraveling a multisystemic illness with unexplained symptoms

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Image in medicine

A 67-year-old Caucasian male was admitted to our hospital due to general fatigue, weight loss, and chronic diarrhea. Physical examination revealed a pale, malnourished man with pedal edema and hypotension. Initial laboratory exams showed renal impairment, nephrotic range proteinuria, and cholestatic jaundice. A thorough investigation of secondary causes of nephrotic syndrome, including computed tomography imaging, endoscopic evaluation, testing for viral and bacterial infections, and immunological assessment. Echocardiography revealed severe concentric ventricular thickening and diastolic dysfunction, along with elevated NT-proBNP levels. Surprisingly, during hospitalization, he

developed unprovoked bilateral periorbital ecchymoses as well as extended ecchymosis after an abdominal wall fat pad biopsy. Meanwhile, plasma and urine immunofixation revealed monoclonal lambda-free light chains with Bence-Jones proteinuria. Biopsies from the petechial mucosa of the ascending colon showed dense amyloid deposits with lambda light chain staining on immunohistochemistry and green birefringence on polarized light microscopy when stained with Congo red dye. Based on the above clinical and immunohistological findings, we made the diagnosis of systemic light chain amyloidosis (AL). Amyloidosis (AL) is a kaleidoscopic disease manifested with a constellation of nonspecific symptoms such as unexplained proteinuria, orthostatic hypotension, gastrointestinal motility issues, peripheral neuropathy with autonomic features, hepatomegaly, cholestatic jaundice, restrictive myocardiopathy, and bleeding diathesis. The spontaneous appearance of periorbital ecchymoses, also known as raccoon eyes sign, is an uncommon but pathognomonic symptom of amyloidosis attributable to increased vascular fragility from the accumulation of amyloid fibrils. Herein, the raccoon eyes sign should prompt internists to include amyloidosis in differential diagnosis to prevent diagnostic delays.

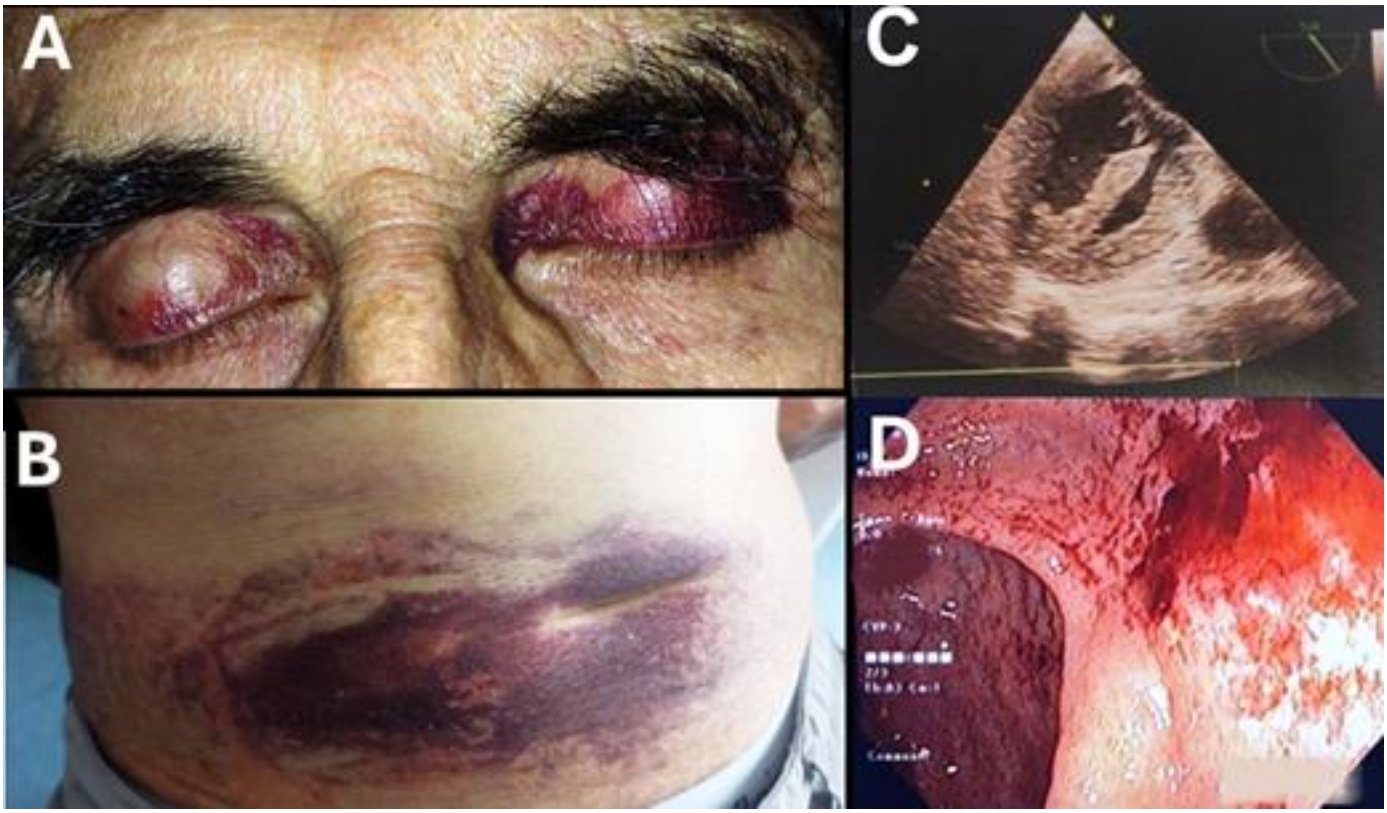

Figure 1: A) spontaneous bilateral periorbital ecchymoses, also known as raccoon eyes sign; B) abdominal wall fat pad biopsy associated extended periumbilical ecchymosis; C) severe concentric ventricular thickening on cardiac ultrasound indicative of restrictive myocardiopathy; D) diffuse frail and petechial mucosa of the ascending colon on endoscopic evaluation