A 60-year-old woman presented with a 1-year history of fatigue, weight loss, polyarthralgia, and non-itchy papules on her eyelids. She also reported paresthesia on hands and feet in the last 4 months. She denied other symptoms. Examination showed multiple symmetric waxy papules on the periorbital region exerting mild mechanical ptosis (Figure). Neurologic examination suggested bilateral carpal tunnel syndrome and loss of sensation on the feet. The remainder physical examination was unremarkable. Laboratory examination was altered for a hemoglobin of 9.0 mg/dL and serum immunoelectrophoresis with monoclonal paraprotein of κ light chain. A skin biopsy specimen of the lesions was positive for Congo red staining with immunohistochemistry confirming the diagnosis of light-chain systemic amyloidosis. Many signs have been described as highly suggestive of light-chain systemic amyloidosis such as shoulder pad sign, macroglossia, and the “raccoon sign.”

We present a rare form of systemic amyloidosis with periorbital involvement characterized by non-itchy waxy papules and alert physicians that this uncommon sign should prompt an extensive workup for occult monoclonal gammopathy and systemic disease.

REFERENCES