Periorbital Papular Amyloidosis

Carlos Antonio Moura, MD,* Marconi Cedro,* and Carlos Geraldo Moura*

A 60-year-old-woman presented with 1-year history of fatigue, weight loss, polyarthralgia, and nonitchy 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



FIGURE. Periorbital papular amyloidosis. Color online figure is available at http://www.jclinrheum.com.

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,¹ macroglossia,² and the "raccoon sign."³ We present a rare form of systemic amyloidosis with periorbital involvement characterized by nonitchy waxy papules⁴ and alert physicians that this uncommon sign should prompt an extensive workup for occult monoclonal gammopathy and systemic disease.

REFERENCES

- 1. Guerreiro de Moura CG, Pinto de Souza SP. Images in clinical medicine. "Shoulder pad" sign. *N Engl J Med.* 2004;351:e23.
- Moura CG, Moura TG, Durães AR, et al. Exuberant macroglossia in a patient with primary systemic amyloidosis. *Clin Exp Rheumatol*. 2005; 23:428.
- de Moura CG, Cruz CM, de Souza SP. Raccoon sign. Arthritis Rheum. 2013; 65:692.
- Kumar S, Sengupta RS, Kakkar N, et al. Skin involvement in primary systemic amyloidosis. *Mediterr J Hematol Infect Dis.* 2013;5:e2013005.

- Correspondence: Carlos Antonio Moura, MD, Hospital Santo Antonio, Salvador, Bahia, Brazil. E-mail: caggmoura@yahoo.com.br.
- Copyright © 2017 Wolters Kluwer Health, Inc. All rights reserved.

From the *Clínica Médica, Hospital Santo Antonio, Salvador, Bahia, Brazil. The authors declare no conflict of interest.

ISŚN: 1076-1608

DOI: 10.1097/RHU.00000000000545