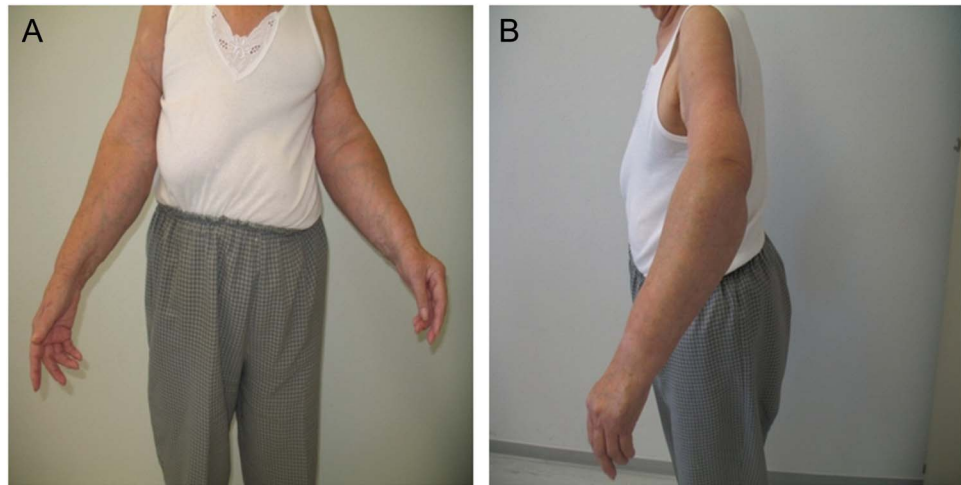


Pseudo-Popeye syndrome

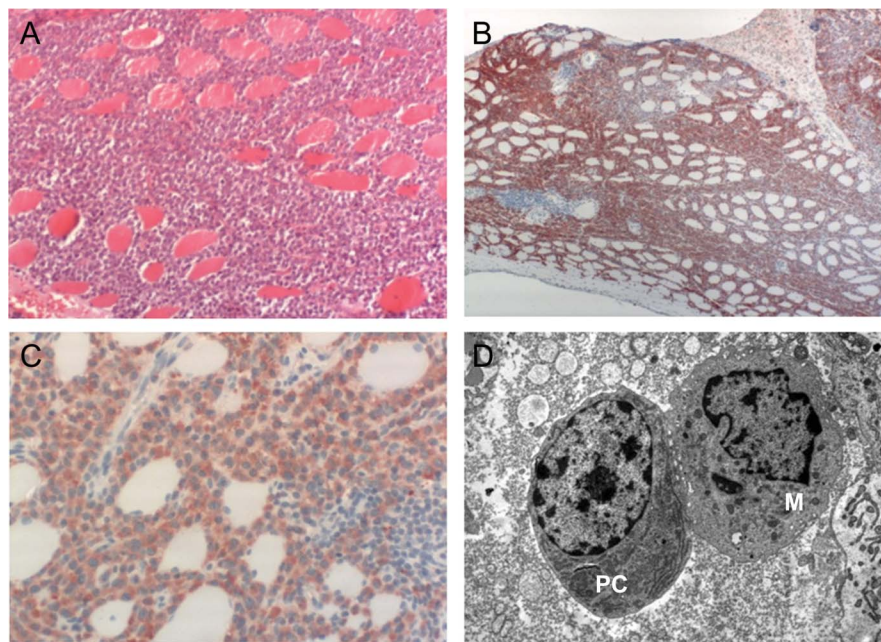
Extramedullary plasmacytoma manifesting in skeletal muscle

Figure 1 Clinical presentation



Front and side views of the patient demonstrate the Popeye effect. The patient complained of painful swelling of both forearms (A), most evident on the left side (B). A massive tumorous bulge caused compartment syndrome, which affected the median and ulnar nerves on the left side.

Figure 2 Histopathologic findings



Muscle biopsy revealed some atrophic muscle fibers displaced by infiltrating immunocytes, visualized using hematoxylin & eosin, 100 \times (A). Immunohistochemical analysis identified plasma cells using CD138 antibody, 50 \times (B) with isolated secretion of kappa light chain, 400 \times (C). Plasma cell character was confirmed ultrastructurally, PC plasma cell, M macrophage (D).

A 75-year-old woman with medullary plasmacytoma reported progressive swelling of the forearms, left more than right (figure 1), with median nerve contribution pain. Examination revealed distal sensory loss and left hand weakness with atrophy of the median and ulnar nerve-innervated muscles. Arterial pulses were palpable. Electrodiagnostic studies showed ulnar nerve conduction block and left thenar muscle denervation. MRI showed subcutaneous and muscle edema in the forearm flexor muscles, mimicking Popeye syndrome.¹ Fasciotomy for compartment syndrome and muscle biopsy were performed. Muscle showed lymphoplasmacytic infiltrates (figure 2), a rare extramedullary manifestation of plasmacytoma.^{2,3} Sensory loss and muscle strength improved after fasciotomy. Therapy for plasmacytoma was bortezomib/dexamethasone plus radiotherapy of both forearms.

Ilka Schneider, MD, Tobias Müller, MD, Gisela Stoltenburg, Professor, Hans-Jürgen Holzhausen, Professor, Christian Mawrin, Professor, Frank Hanisch, MD

From Martin-Luther-University Halle-Wittenberg (I.S., T.M., G.S., H.-J.H., F.H.), Halle (Saale); Institute of Cell and Neurobiology (G.S.), Charité, Universitätsmedizin Berlin; and Institute of Neuropathology (C.M.), Otto-von-Guericke-University Magdeburg, Germany.

Author contributions: Ilka Schneider: analysis and interpretation of the data, histologic and immunohistologic analysis of the muscle biopsy. Tobias Müller: providing pictures of the patient, performing muscle biopsy, revising the manuscript for intellectual content. Gisela Stoltenburg: histologic and immunohistologic analysis of the muscle biopsy, revising the manuscript for intellectual content. Hans-Jürgen Holzhausen: performing ultrastructural analysis of the muscle biopsy, revising the manuscript for intellectual content. Christian Mawrin: performing histologic and immunohistologic staining on formalin-fixed muscle biopsy. Frank Hanisch: conceptualization of the study, revising the manuscript for intellectual content, histologic and immunohistologic analysis of the muscle biopsy.

Study funding: No targeted funding reported.

Disclosure: I. Schneider received compensation for travel expenses and accommodation from Genzyme Corporation in 2012. T. Müller received honoraria for lectures from Boehringer-Ingelheim and Bayer AG. F. Hanisch received lecturer honoraria and travel fees from Genzyme, Astellas, and Biomarin Incorp. G. Stoltenburg, H. Holzhausen, and C. Mawrin report no disclosures. Go to Neurology.org for full disclosures.

Correspondence to Dr. Schneider: ilka.schneider@medizin.uni-halle.de

1. Biemans RG. The Popeye syndrome: brachial artery entrapment as a result of muscular hypertrophy. *Neth J Surg* 1984;36:103–106.
2. Sekiguchi Y, Asahina T, Shimada A, et al. A case of extramedullary plasmablastic plasmacytoma successfully treated using a combination of thalidomide and dexamethasone and a review of the medical literature. *J Clin Exp Hematop* 2013;53:21–28.
3. Thoumazet F, Donnio A, Ayeboua L, Brebion A, Diedhou A, Merle H. Orbital and muscle involvement in multiple myeloma. *Can J Ophthalmol* 2006;41:733–736.

Neurology®

Pseudo-Popeye syndrome: Extramedullary plasmacytoma manifesting in skeletal muscle

Ilka Schneider, Tobias Müller, Gisela Stoltenburg, et al.

Neurology 2014;82;544-545

DOI 10.1212/WNL.0000000000000099

This information is current as of February 10, 2014

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/82/6/544.full
References	This article cites 3 articles, 0 of which you can access for free at: http://n.neurology.org/content/82/6/544.full#ref-list-1
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): All Immunology http://n.neurology.org/cgi/collection/all_immunology All Neuromuscular Disease http://n.neurology.org/cgi/collection/all_neuromuscular_disease Hematologic http://n.neurology.org/cgi/collection/hematologic Muscle disease http://n.neurology.org/cgi/collection/muscle_disease
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2014 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

