Immunogammopathies and Acquired Vitelliform Detachments: A Report of Four Cases

IRENE M. RUSU, SARAH MREJEN, MICHAEL ENGELBERT, ROBERTO GALLEGO-PINAZO, MICHAEL D. OBER, MARK W. JOHNSON, ANITA LEYS, AND LAWRENCE A. YANNUZZI

• PURPOSE: To describe the nature and evolution of acquired macular detachments in patients with immunogammopathies and to propose a mechanism for their development.

• DESIGN: Retrospective observational case series.

• METHODS: Three patients with multiple myeloma and 1 with light chain deposition disease were diagnosed with vitelliform macular detachments based on clinical examination, fundus autofluorescence, fluorescein angiography, and optical coherence tomography. These patients were followed over time and their clinical examinations and imaging studies were compared and contrasted.

• RESULTS: Three patients (5 eyes) with multiple myeloma and 1 patient (2 eyes) with light chain deposition disease presented with acquired macular yellowish subretinal deposits on funduscopic examination that corresponded to hyperautofluorescent lesions on fundus autofluorescence imaging and subretinal hyperreflective material on spectral-domain optical coherence tomography. One patient (2 eyes) had diffuse serous retinal detachments involving not only the macular region but also the midperiphery of the retina. These acquired macular vitelliform detachments were not associated with signs of hyperviscosity retinopathy in 5 eyes and resolved after successful treatment of the multiple myeloma in 6 eyes.

• CONCLUSION: Patients with an immunogammopathy such as multiple myeloma or light chain deposition disease may develop serous elevations of the macula that we classify as acquired vitelliform detachments using multimodal imaging. Appropriate evaluation including serum protein electrophoresis and hematology consultation should be considered in the management of patients with acquired vitelliform detachments of uncertain etiology. (Am J Ophthalmol 2014;157:648–657. © 2014 by Elsevier Inc. All rights reserved.)

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From the Department of Ophthalmology, New York University School of Medicine, New York, New York (I.M.R., M.E., L.A.Y.); Vitreous Retina Macula Consultants of New York, New York, New York (S.M., M.E., L.A.Y.); LuEsther T. Mertz Retinal Research Center, Manhattan Eye, Ear and Throat Hospital-Northshore Long Island Jewish Hospital, New York, New York (S.M., M.E., L.A.Y.); Department of Ophthalmology, University and Polytechnic Hospital La Fe, Valencia, Spain (R.G.P.); Edward S. Harkness Eye Institute, Columbia University College of Physicians and Surgeons, New York, New York (M.E., L.A.Y.); Retina Consultants of Michigan, Southfield, Michigan (M.D.O.); Kellogg Eye Center, University of Michigan, Ann Arbor, Michigan (M.W.J.); and University Hospitals Leuven, Leuven, Belgium (A.L.).

Inquiries to Lawrence A. Yannuzzi, Vitreous Retina Macula Consultants of New York, 460 Park Avenue, Fifth Floor, New York, NY 10022; e-mail: layannuzzi@gmail.com

IMMUNOGAMMOPATHIES ARE CLONAL PLASMA CELL proliferative disorders characterized by deposition of light or heavy chain immunoglobulin fragments in tissues, leading to organ dysfunction. Within this spectrum of diseases are multiple myeloma, Waldenstrom's macroglobulinemia, and benign monoclonal gammopathy, as well as light chain deposition disease. Ocular manifestations of immunogammopathies have been described in a variety of ocular structures, including the conjunctiva, cornea, uvea, and retina.[1]

Serous macular detachments in association with immunogammopathies, though rare, have been described.[2–15] Ho and associates first reported serous macular detachments with or without subretinal precipitates or fundus signs of serum hyperviscosity in 1 patient with multiple myeloma, 1 patient with Waldenstrom's macroglobulinemia, and 1 patient with benign polyclonal gammopathy.[2] Specifically in multiple myeloma, only 1 report describes deposits on the posterior surface of the neurosensory retina and in the subretinal space anterior to the retinal pigment epithelium (RPE).[2] This previous study did not include current imaging modalities such as spectral-domain optical coherence tomography (SD OCT) or fundus autofluorescence imaging.

This is a report on an additional 4 patients—3 with multiple myeloma and 1 with light chain deposition disease—who have associated serous detachments evaluated with multimodal imaging including fluorescein angiography, fundus autofluorescence, and SD OCT. We also propose a mechanism that would explain the advent and course of these particular exudative detachments.

PATIENTS AND METHODS

THE INSTITUTIONAL REVIEW BOARD (IRB) AT NORTHSHORE Long Island Jewish Hospital waived IRB approval for this retrospective case series. The records of 4 patients with vitelliform macular detachments and an immunogammopathy were examined. The following data were collected for each of the 4 cases: Snellen visual acuity at presentation

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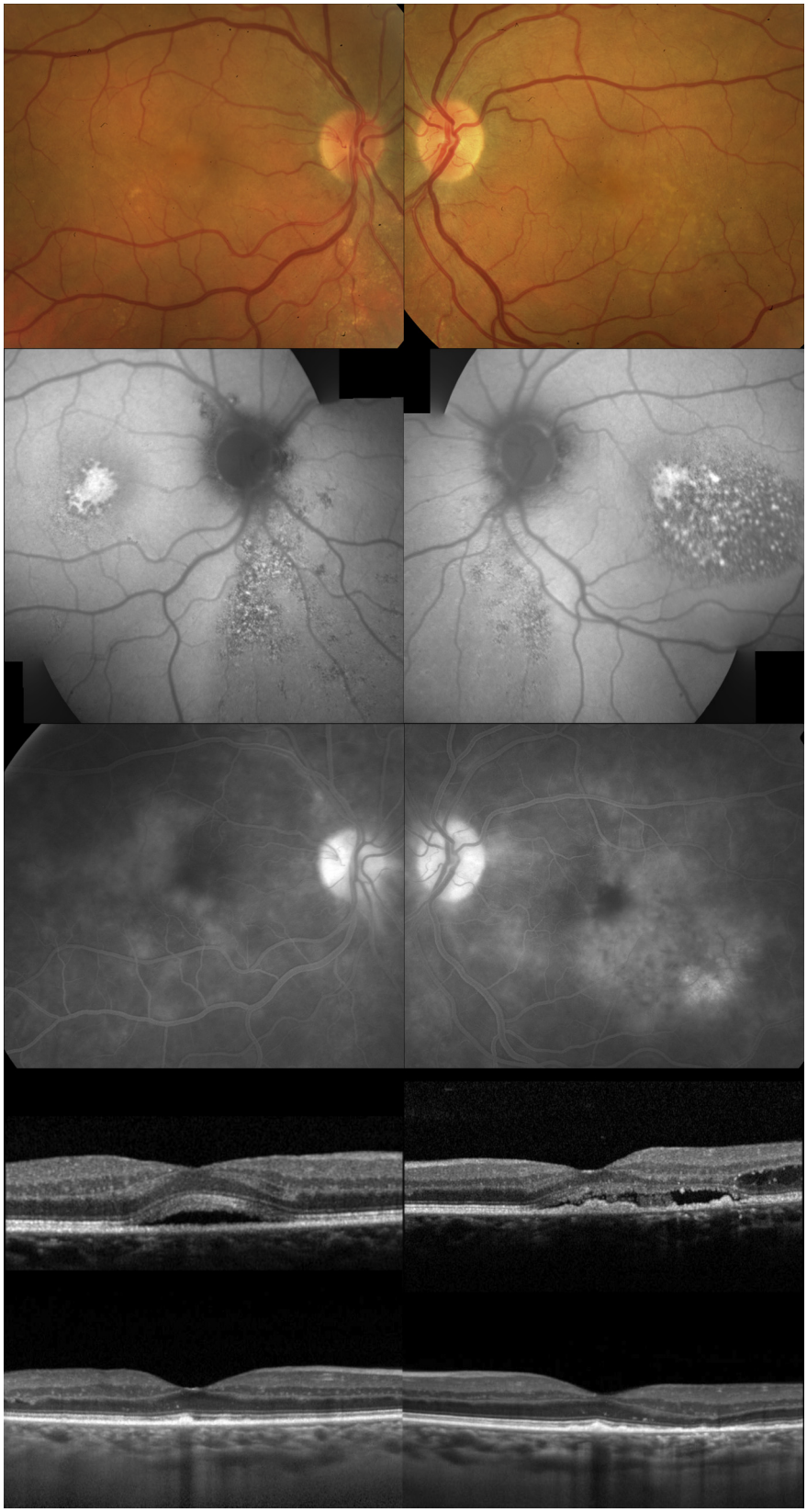

FIGURE 1. Multimodal imaging of both eyes of a 58-year-old woman with multiple myeloma presenting with atrophic descending tracts and bilateral acquired vitelliform detachments that resolved when the serum immunoglobulin levels decreased. (Top row) Fundus photographs show mild retinal pigment epithelium changes. (Second row) Fundus autofluorescence shows hyperautofluorescent macular lesions surrounded by a wider area of granular hypoautofluorescence, more prominent OS, and bilateral atrophic