

serving on a Scientific Advisory or Data Safety Monitoring board for Alexion. The institution of Dr. Pittock has received research support from Grifols. The institution of Dr. Pittock has received research support from NIH. The institution of Dr. Pittock has received research support from Viela Bio/MedImmune/Horizon. The institution of Dr. Pittock has received research support from Alexion Pharmaceuticals. The institution of Dr. Pittock has received research support from F. Hoffman/LaRoche/Genentech. Dr. Pittock has received intellectual property interests from a discovery or technology relating to health care. Dr. Pittock has received intellectual property interests from a discovery or technology relating to health care. The institution of Dr. McKeon has received research support from Euroimmun AG. The institution of Dr. McKeon has received research support from National Institutes of Health. Dr. McKeon has received intellectual property interests from a discovery or technology relating to health care. Dr. McKeon has received intellectual property interests from a discovery or technology relating to health care. Dr. McKeon has received publishing royalties from a publication relating to health care. Dr. Klein has a non-compensated relationship as a Klein with Neurology Journal that is relevant to AAN interests or activities. The institution of Dr. Dubey has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for UCB. The institution of Dr. Dubey has received personal compensation in the range of \$5,000-\$9,999 for serving as a Consultant for Astellas. Dr. Dubey has received personal compensation in the range of \$0-\$499 for serving on a Speakers Bureau for AGRIMS. Dr. Dubey has received personal compensation in the range of \$0-\$499 for serving on a Speakers Bureau for Advances in Neurology. Dr. Dubey has received personal compensation in the range of \$0-\$499 for serving on a Speakers Bureau for Moffit Cancer Center. Dr. Dubey has received research support from Department of Defense. Dr. Dubey has received intellectual property interests from a discovery or technology relating to health care. Dr. Dubey has received intellectual property interests from a discovery or technology relating to health care. Dr. Mills has received intellectual property interests from a discovery or technology relating to health care.

CAR-T Cell-Mediated B Cell Depletion in Central Nervous System Autoimmunity

Sasha Gupta, Milos Simic, Sharon Sagan, Jason Duecker, Channele Shepherd, Raymond Sobel, Stephen Hauser, Wendell Lim, Michael Wilson, Scott Zamvil

Objective

Evaluate chimeric antigen receptor (CAR)-T cell mediated B cell depletion in experimental autoimmune encephalomyelitis (EAE).

Background

CAR-T cells are autologous T cells expressing a non-MHC target antigen specific receptor. We tested whether anti-CD19 CAR-T cells, which more thoroughly deplete human B cell populations than monoclonal antibodies (mAbs), recapitulated the beneficial effects of B cell depletion in EAE.

Design/Methods

Anti-CD19 CAR-T cells or control T cells that overexpressed green fluorescent protein were transferred into female wild-type C57BL/6 mice that had been pretreated with cyclophosphamide. EAE was induced by immunization with either recombinant human (rh) myelin oligodendrocyte protein (MOG) (B cell-dependent) or MOG peptide (p) 35-55 (B cell-independent). Mice were evaluated daily for clinical signs of EAE and weekly for peripheral B and T cell counts. B cell levels, T cell immune modulation and histology were assessed at peak disease and at termination.

Results

In rhMOG-induced EAE, clinical scores and histologic lymphocyte infiltration were reduced in mice treated with cyclophosphamide and either anti-CD19 CAR-T cells or control T cells. B cell depletion was observed in peripheral lymphoid tissue and in the central nervous system (CNS) of mice treated with anti-CD19 CAR T cells, similar to effects of anti-CD20 mAbs. There was no difference in T cell modulation including Th1 or Th17 populations, but there was a trend towards increase in Treg populations in the periphery and CNS in the anti-CD19

CAR-T cell and control T cell treated animals. Clinical scores and histology did not differ among treatment groups in p35-55-induced disease.

Conclusions

Anti-CD19 CAR-T cells thoroughly deplete B cells peripherally and within the CNS. Treatment also results in less severe rhMOG-induced disease, but it was independent of B cell depletion. Our results are consistent with human data indicating that anti-CD19 CAR-T cells deplete B cells across compartments, suggesting that they may hold promise for progressive MS.

Disclosure: Dr. Gupta has nothing to disclose. Milos Simic has received research support from UCSF and NMSS. Milos Simic has received intellectual property interests from a discovery or technology relating to health care. Sharon Sagan has nothing to disclose. Mr. Duecker has nothing to disclose. Ms. Shepherd has nothing to disclose. Raymond Sobel has a non-compensated relationship as a Deputy Editor with Journal of Neuropathology and Experimental Neurology that is relevant to AAN interests or activities. Dr. Hauser has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for NGM Bio. Dr. Hauser has received personal compensation in the range of \$0-\$499 for serving on a Scientific Advisory or Data Safety Monitoring board for Accure. Dr. Hauser has received personal compensation in the range of \$0-\$499 for serving on a Scientific Advisory or Data Safety Monitoring board for Alector. Dr. Hauser has received personal compensation in the range of \$0-\$499 for serving on a Scientific Advisory or Data Safety Monitoring board for Annexon. Dr. Hauser has received personal compensation in the range of \$10,000-\$49,999 for serving as an officer or member of the Board of Directors for Neurona. Dr. Hauser has a non-compensated relationship as a Clinical Trial/Primary Investigator with Roche that is relevant to AAN interests or activities. Dr. Hauser has a non-compensated relationship as a Clinical Trial/Primary Investigator with Novartis that is relevant to AAN interests or activities. Wendell Lim has nothing to disclose. Dr. Wilson has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for WebMD. Dr. Wilson has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Genentech. Dr. Wilson has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Novartis. Dr. Wilson has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Takeda. The institution of Dr. Wilson has received research support from Genentech / Roche. The institution of Dr. Wilson has received research support from NIH. The institution of Dr. Wilson has received research support from UCSF Weill Institute for Neurosciences. The institution of Dr. Wilson has received research support from Novartis. Dr. Wilson has received personal compensation in the range of \$5,000-\$9,999 for serving as a Expert Witness with US Dept of Justice. Dr. Zamvil has received personal compensation in the range of \$500-\$4,999 for serving on a Speakers Bureau for Alexion. Dr. Zamvil has received personal compensation in the range of \$500-\$4,999 for serving on a Speakers Bureau for Genzyme. Dr. Zamvil has received personal compensation in the range of \$500-\$4,999 for serving as an Editor, Associate Editor, or Editorial Advisory Board Member for AAN. Dr. Zamvil has received personal compensation in the range of \$500-\$4,999 for serving as a Advisory Board with Genzyme. Dr. Zamvil has received personal compensation in the range of \$500-\$4,999 for serving as a Advisory Board with Genentech. Dr. Zamvil has received personal compensation in the range of \$500-\$4,999 for serving as a Advisory Board with Alexion.

A Rare Neuromyelitis Optica Mimic: Primary CNS Histiocytic Sarcoma

David Rogawski, Jeffrey Nirschl, Jamie McDonald, Esther Nie, Nicholas Schwartz, Hannes Vogel, Brian Scott, Carl Gold, Lucas Kipp

Objective

NA.

Background

NA.

Design/Methods

NA.

Results

Primary CNS histiocytic sarcoma is a rare hematolymphoid malignancy with features of mature histiocytes and carries a poor prognosis. We describe a unique case in which a 50-year-old woman presented with recurrent acute brainstem syndrome, area postrema syndrome, and myelitis with corresponding MRI lesions meeting diagnostic criteria for seronegative NMOSD. Despite initial improvement with steroids and plasma exchange, she experienced recurrent symptoms over ten months referable to new and persistently enhancing lesions. At autopsy, neuropathology revealed a diffusely infiltrative primary CNS histiocytic sarcoma. This case represents a rare clinicoradiologic mimic of NMOSD, underscoring the importance of evaluation for infiltrative diseases in cases of atypical seronegative NMOSD.

Conclusions

NA.

Disclosure: Dr. Rogawski has nothing to disclose. Mr. Nirschl has received intellectual property interests from a discovery or technology relating to health care. Dr. McDonald has nothing to disclose. An immediate family member of Dr. Nie has received personal compensation for serving as an employee of Collective Acumen. Dr. Schwartz has nothing to disclose. Dr. Vogel has received personal compensation in the range of \$500-\$4,999 for serving as an Expert Witness for Lipton Law. Dr. Vogel has received publishing royalties from a publication relating to health care. Dr. Scott has nothing to disclose. Dr. Gold has received personal compensation for serving as an employee of Stanford University. An immediate family member of Dr. Gold has received personal compensation for serving as an employee of Stanford University. The institution of Dr. Kipp has received research support from Biogen. The institution of Dr. Kipp has received research support from Genentech. The institution of Dr. Kipp has received research support from Sanofi-Genzyme.

Recurrent Foreign Body Reactions to Neuroendovascular Polymers—A Clinicopathologic Case Study

William Lou, Thuhien Nguyen, Caitlin Latimer

Objective

We provide histopathologic and neuroimaging evidence of recurrent foreign body reactions in a patient following separate stent-assisted coiling of two contralateral intracranial aneurysms a decade apart.

Background

Stent-assisted endovascular coiling for wide-neck cerebral aneurysms introduces permanent foreign materials into the cerebral vasculature. While foreign body reactions after endovascular coiling are increasingly reported in the literature, compelling histopathologic data remains very limited.

Design/Methods

Electronic medical record review for clinical details and neuroimaging. Histopathology was extensively reviewed with a neuropathologist.

Results

A 37-year-old woman presents with left arm weakness. Magnetic resonance imaging (MRI) shows numerous enhancing lesions with large multifocal T2 FLAIR changes in the right hemisphere. An extensive vascular, infectious, autoimmune, and neoplastic workup returns negative. Months earlier, she had undergone stent-assisted coiling of a right internal carotid artery (ICA) aneurysm. A decade prior, she had presented with focal right-sided seizures after coiling of a left ICA aneurysm; MRI brain at the time revealed two enhancing lesions in the left hemisphere of unclear etiology. A brain biopsy is performed, and histopathology reveals multifocal, chronic micro-abscesses characterized by collections of neutrophils surrounded by a rim of multinucleated giant cells and histiocytes which are in turn rimmed by fibrosis and granulation tissue. Staining is negative for neoplastic changes and infectious organisms. Rare filamentous structures are identified in association with the giant cells; these resemble coil polymers described in the endovascular literature and are highly suspicious for inducing a neuroinflammatory foreign body reaction. She

improves following glucocorticoid treatment, and repeat imaging shows substantial reduction in parenchymal abnormalities.

Conclusions

Foreign body reactions are an uncommon complication of endovascular aneurysm coiling and can manifest as an embolic inflammatory phenomenon. These neuroinflammatory reactions are driven by endovascular polymers and responsive to glucocorticoid treatment. Heightened awareness can facilitate earlier diagnosis and treatment, with prompt neuroimmunology consultation for repeat endovascular procedures.

Disclosure: Dr. Lou has nothing to disclose. An immediate family member of Dr. Nguyen has received personal compensation for serving as an employee of Caption Health. An immediate family member of Dr. Nguyen has stock in Caption Health. An immediate family member of Dr. Nguyen has received intellectual property interests from a discovery or technology relating to health care. The institution of Dr. Latimer has received research support from NIA. Dr. Latimer has received publishing royalties from a publication relating to health care.

Neuro-Behcet's Disease Presenting as a Pseudotumoral Brainstem Mass: A Case Report

Heather Yong, Carlos Camara-Lemarrroy, Katayoun Alikhani

Objective

Herein, we present a rare and diagnostically challenging case of neuro-Behcet disease (NBD) manifesting as a pseudotumoral brainstem mass.

Background

Pseudotumoral-NBD as a neurologic manifestation of Behcet's disease is rare. Imaging is characterized by mass-like lesions that enhance with contrast, are hyperintense with T2-weighted and fluid-attenuated inversion recovery (FLAIR) and show restricted diffusion. The differential includes glial lesions, lymphomas, infectious and granulomatous lesions.

Design/Methods

This is a case study of a 33-year-old male of West-African descent with a history of Behcet's disease.

Results

A 33-year-old male of West-African descent with a history of Behcet's disease, presented two years after his diagnosis with headaches, low-grade fever, genital ulcerations, and horizontal binocular diplopia. Imaging revealed a large right-sided T2/FLAIR hyperintense abnormality in the medulla with a central area of necrosis. Cerebrospinal fluid revealed lymphocytic-predominant pleocytosis with 11·10⁶ cells/L (reference range 0-5), and high levels of interleukin-6. His vasculitis, infectious, paraneoplastic, flow cytometry, and autoimmune panels were negative. He tested positive for hepatitis-B core antigen, and latent tuberculosis. The etiology for his presentation was believed to be parenchymal NBD and he received a 3-day course of intravenous solumedrol (eventually transitioned to prednisone and azathioprine) with significant improvement. Imaging 1-week post-treatment revealed resolution of enhancement, and at 3 months he had near complete lesion resolution.

Conclusions

NBD can rarely present with a pseudotumoral presentation, which can cause diagnostic uncertainty. A thorough radiologic/laboratory workup should be conducted to exclude other neurologic diagnoses; however, a high index of suspicion for NBD is required in similar cases and a spectacular response to steroids are invaluable in diagnosis.

Disclosure: Dr. Yong has nothing to disclose. Dr. Camara-Lemarrroy has nothing to disclose. Dr. Alikhani has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Apotex, Biogen, Bristol Myers Squibb, EMD Serono, Novartis, Roche, Sanofi Genzyme.

Neurology®

CAR-T Cell-Mediated B Cell Depletion in Central Nervous System Autoimmunity

Sasha Gupta, Milos Simic, Sharon Sagan, et al.

Neurology 2022;99;S32

DOI 10.1212/01.wnl.0000903276.26170.3f

This information is current as of December 5, 2022

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/99/23_Supplement_2/S32.1.full
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): Cerebrospinal Fluid http://n.neurology.org/cgi/collection/cerebrospinal_fluid CT http://n.neurology.org/cgi/collection/ct Low pressure syndrome http://n.neurology.org/cgi/collection/low_pressure_syndrome
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 © 2022 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

