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.

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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.

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

Heather Yong, Carlos Camara-Lemarroy, Katayoun Alikhani

Objective

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

Background

Psuedotumoral-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 psuedotumoral 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.

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