The Eye as a Window to the Brain: Prominent Retinal Vasculopathy Points to Neuro-Behcet Diagnosis for an Undifferentiated Solitary Brain Lesion

Sonam Dilwali, Adil Harroud, Nailyn Rasool, Ari Green

Objective

To report a perplexing case of Behcet disease (BD) presenting as a focal parenchymal lesion that reached a diagnosis after noting a prominent retinal vasculopathy, highlighting the importance of ophthalmologic evaluation in undifferentiated CNS disease.

Background

BD can have variable systemic manifestations driven by a vasculitis, including oral or genital ulcers, pulmonary aneurysms, and uveitis. Neurologic involvement is present in less than 10% of patients, most commonly as a meningoencephalitis.

Design/Methods

We present the case of a woman who developed asymmetric sub-acute sensorineural hearing loss at age 31 followed by transient right facial weakness at age 40, and most recently presented with right facial numbness and arm weakness at age 47. Brain MRI revealed a left frontal enhancing lesion with associated T2/FLAIR hyperintensity extending from the periventricular to the juxtacortical area with a thin rim of a reduced diffusion. CSF and serum studies were negative for inflammation, infection and malignancy except for elevated ESR and CRP. Brain biopsy revealed non-specific gliosis. Persistent enhancement on MRI was noted over 3 months, with spontaneous clinical improvement. Patient endorsed insidious vision changes over recent years, and visual testing was performed.

Results

Dilated ophthalmic examination demonstrated striking peripheral attenuation and sclerosis of retinal vasculature, with evidence of non-perfusion and skip lesions on retinal fluorescein angiography (FA). Findings of occlusive retinal peripheral vasculopathy suggested an underlying vasculitis as the etiology of the brain lesion and prior deficits, raising the likelihood of BD. Patient was homozygous for HLA-B*51, further supporting this diagnosis even with lack of mucosal ulcers and negative history of pathergy.

Conclusions

Neurologic manifestations of BD can be diverse including retinal occlusive vasculopathy; ulcers are not universally present. Ophthalmologic examination, even when minimally symptomatic, can inform the diagnosis of CNS lesions. Patient was started on Prednisone, Infliximab and Methotrexate, achieving disease remission.

Disclosure: Dr. Dilwali has nothing to disclose. The institution of Dr. Harroud has received research support from National Multiple Sclerosis Society. The institution of Dr. Harroud has received research support from Multiple Sclerosis Society of Canada. Dr. Rasool has nothing to disclose. Dr. Green has received personal compensation in the range of $50,000-$99,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Pipeline Therapeutics. Dr. Green has received personal compensation in the range of $0-$499 for serving on a Scientific Advisory or Data Safety Monitoring board for Bionure. Dr. Green has received personal compensation in the range of $10,000-$49,999 for serving as an Editor, Associate Editor, or Editorial Advisory Board Member for JAMA Neurology. The institution of Dr. Green has received research support from NINDS. The institution of Dr. Green has received research support from NMSS. The institution of Dr. Green has received research support from NIA. The institution of Dr. Green has received research support from Adelson Research Foundation. Dr. Green has received intellectual property interests from a discovery or technology relating to health care. Dr. Green has received personal compensation in the range of $500-$4,999 for serving as a Study Section with NINDS. Dr. Green has a non-compensated relationship as a Author with Viela Bio that is relevant to AAN interests or activities.

A Case of Recurrent Idiopathic Hypertrophic Pachymeningitis After Years of Quiescence

Benjamin Bird, Zahir Sheikh, Jikku Jose Zachariah

Objective

To report a case of idiopathic hypertrophic pachymeningitis with recurrence in a new region of the brain after years of quiescence.

Background

Idiopathic hypertrophic pachymeningitis (IHP) is a rare condition defined by thickening of the dural layer secondary to inflammation without discernible cause. Common symptoms include headache, cranial neuropathies, visual loss, mastoiditis and hearing loss. We present a case of a woman with two discrete episodes of headache and vision changes associated with dural thickening and parenchymal edema in separate locations, eventually with biopsy-supported diagnosis of IHP. A 41-year-old woman presented to our hospital with days of persistent temporal headache, blurred vision and confusion. MRI of the brain with contrast demonstrated left temporal lobe edema and overlying dural thickening, initially concerning for mastoiditis versus malignancy. Bloodwork revealed mildly elevated CRP and chronic untreated hepatitis C (HCV). Lumbar puncture was unrevealing, including cell counts, flow cytometry, cytology, cultures, CSF RPR and herpes simplex. Additional infectious workup, including for tuberculosis and fungi, was negative. IgG4 levels were normal, and ANCA screening was negative. CT of the chest revealed lung and liver nodules with non-specific inflammation on biopsy. Mastoidectomy with myringotomy showed no infection. PET scan was unremarkable. Ultimately, biopsy of dural thickening showed chronic inflammation, predominantly CD-163+ histiocytes without granulomas or malignancy. Seven years prior, the patient suffered a similar episode, with MRI showing extensive bilateral frontal dural thickening with associated edema. Symptoms resolved after course of corticosteroids with taper, though minor right frontopolar gliosis persisted. IHP suspected after similar workup, but no biopsy performed.

Design/Methods

NA.

Results

NA.

Conclusions

IHP is clinically well-described, but data on course and recurrence patterns over time is scarce. This case demonstrates that recurrence is not restricted to original affected areas and can happen after years of quiescence. Given course and positive HCV, further longitudinal follow-up and studies are warranted.

Disclosure: Dr. Bird has nothing to disclose. Dr. Sheikh has nothing to disclose. Dr. Zachariah has nothing to disclose.

Unmasking of a Relapsing Encephalomyelitis After SARS-CoV-2 Infection and COVID-19 Vaccination

Shuvro Roy, Paula Barreras, Carlos Pardo-Villamizar, Scott Newsome

Objective

NA.

Background

Prior case studies suggest that severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) and its vaccines may unmask neuroinflammatory conditions. We present a case of relapsing steroid-responsive encephalomyelitis after SARS-CoV-2 infection and subsequent COVID-19 vaccination.

Design/Methods

NA.

Results

A 47-year-old man with a history COVID-19 presented with subacute lower extremity weakness, erectile dysfunction, and gait instability with falls. His symptoms started several weeks after COVID-19 vaccination which he underwent 3 months after COVID-19 infection. His initial exam demonstrated weakness at the knees and ankles and extensor plantar responses. MRI demonstrated innumerable enhancing lesions involving the subcortical white matter, basal ganglia, thalami, brainstem, cerebellum, and the entire spinal cord parenchyma. CSF testing revealed a lymphocytic pleocytosis (10 WBC, 88% lymphocytes), and transient matched serum and CSF oligoclonal bands. Testing was unremarkable for infections, malignancies, primary demyelinating conditions, etc. He responded dramatically to five days of high dose methylprednisolone but had recurrence of symptoms with weaning of oral prednisone, requiring another pulse of IV steroids. After 2 months, his steroids were weaned again, with clinical and radiographic recurrence, requiring another course of IV steroids. He was subsequently transitioned to mycophenolate as a steroid-sparing agent. Literature review identified 20 additional cases of CNS neuroinflammatory disease after either SARS-CoV-2 infection or vaccination (11 transverse myelitis, 6 optic neuritis, 3 encephalomyelitis).

Conclusions

Our patient's steroid-dependency and relapsing course suggests unmasking of an underlying CNS neuroinflammatory condition. Temporal associations of neurological conditions with vaccinations or infections do not prove causality despite previous reports of such sequelae. Vaccines containing SARS-CoV-2 antigens may enhance autoimmunity by mechanisms including polyclonal activation, epitope spreading, or molecular mimicry. This case highlights that the resulting inflammation may be insidious and extensive, though treatable. As COVID-19 constitutes a life-threatening infection in some patients, the benefits of vaccination outweigh the smaller risk of unmasking an immune-related condition.

Disclosure: Dr. Roy has nothing to disclose. The institution of Dr. Barreras has received research support from Foundation for Sarcoidosis Research. The institution of Dr. Pardo-Villamizar has received research support from National Institutes of Health. The institution of Dr. Pardo-Villamizar has received research support from Bart McLean Fund for Neuroimmunology Research. Dr. Newsome has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Autobahn. Dr. Newsome has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Biogen. Dr. Newsome has received personal compensation in the range of $10,000-$49,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Genentech. Dr. Newsome has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Novartis. Dr. Newsome has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for EMD Serono. Dr. Newsome has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Greenwich Biosciences. Dr. Newsome has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Bristol Myers Squibb. Dr. Newsome has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Horizon Therapeutics. The institution of Dr. Newsome has received research support from Biogen. The institution of Dr. Newsome has received research support from Genentech/Roche. The institution of Dr. Newsome has received research support from Department of Defense. The institution of Dr. Newsome has received research support from Patient Centered Outcomes Research Institute. The institution of Dr. Newsome has received research support from National MS Society. Dr. Newsome has received personal compensation in the range of $5,000-$9,999 for serving as a Clinical adjudication committee member for clinical trial with medDay Pharmaceuticals. Dr. Newsome has received personal compensation in the range of $10,000-$49,999 for serving as a Lead PI for Clinical Trial with Roche.

Characteristics and Evolution of Cerebral Aneurysms Among Adults Living With HIV: A Retrospective, Longitudinal Case Series

Emily I White, Pria Anand, Anna Marisa Cervantes-Arslanian

Objective

To describe the characteristics and evolution of cerebral aneurysms in a large cohort of adults living with human immunodeficiency virus (ALWH).

Background

HIV-associated vasculopathy, which can predispose to cerebral aneurysm development, may result from an inflammatory process whereby HIV viral proteins lead to increased chemoattractant and adhesion particles in the vessel wall. Additionally, adventitial layer inflammation has been shown to be strongly associated with HIV infection and vascular changes that may lead to cerebral aneurysm formation. A recent population-based matched cohort study demonstrated a higher rate of subarachnoid hemorrhage in ALWH. It is important to expand the current understanding of risk factors for aneurysm development and the longitudinal course of patients with HIV-associated vasculopathy to help reduce this source of potential morbidity.

Design/Methods

The Clinical Data Warehouse was queried for all adult patients evaluated at Boston Medical Center between January 1, 2000, and October 22, 2021 with a history of HIV and at least one cerebral aneurysm. Charts were reviewed for variables including timing of HIV and aneurysm diagnoses, antiretroviral (ART) duration, additional aneurysm risk factors, development of new aneurysms/growth of existing aneurysms, interventions, and clinical outcomes.

Results

A total of 50 patients (52% female) were identified, including 82 cerebral aneurysms. Forty-six percent of patients with a nadir CD4 count less than 200 cells/mm^3 (N = 13) developed new aneurysms or were found to have aneurysm growth over time compared with 29% of patients with a CD4 nadir above 200 cells/mm^3 (N = 21). New aneurysms were found or existing aneurysms grew in 67% of those not on ART at time of aneurysm diagnosis (N = 6), 38% of those with inconsistent ART use (N = 8), and 21% of those with consistent ART (N = 19).

Conclusions

Among ALWH, lower CD4 nadir and inconsistent ART use may contribute to aneurysm formation or growth. Further studies are needed to more thoroughly characterize this trend.

Disclosure: Dr. White has nothing to disclose. Dr. Anand has nothing to disclose. Dr. Cervantes-Arslanian has nothing to disclose.

Headache as Initial Presentation of Human Chronic Necrotizing Granulomatous Meningoencephalitis

Sohaib Lateef, Khaled Gharaibeh, Danya Zamir, Ajaz Sheikh

Objective

Chronic necrotizing and granulomatous meningoencephalitis is an idiopathic inflammatory disease with possible autoimmune mediated delayed type hypersensitivity response. It commonly affects the central nervous system of dogs and cats. However, this inflammatory disease has rarely been described in humans.

Background

A 69-year-old right-handed woman presented with worsening subacute headaches that were intermittent, sharp and holocephalic in nature which worsened with coughing and/or laughing. Her initial neurological

Unmasking of a Relapsing Encephalomyelitis After SARS-CoV-2 Infection and COVID-19 Vaccination

Shuvro Roy, Paula Barreras, Carlos Pardo-Villamizar, et al.

Neurology 2022;99;S61-S62
DOI 10.1212/01.wnl.0000903492.27780.7c