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 discernable 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.

Results

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.

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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 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 $10,000-$49,999 for serving as an Advisory or Data Safety Monitoring board for Pipeline Therapeutics. 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 $50,000-$99,999 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 Associate 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 an Author with Viela Bio that is relevant to AAN interests or activities.

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Unmasking of a Relapsing Encephalomyelitis After SARS-CoV-2 Infection and COVID-19 Vaccination
Shuvro Roy, Paula Barreras, Carlos Pardo-Villamizar, Scott Newsome

Objective
To report a case of relapsing steroid-responsive encephalomyelitis after SARS-CoV-2 infection and subsequent COVID-19 vaccination.

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

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