

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.

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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/mm³ (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/mm³ (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

examination was unremarkable. MRI of the brain revealed multifocal enhancing lesions and edema as well as multifocal signal abnormality throughout the supratentorial and infratentorial parenchyma. Her cerebrospinal fluid showed elevated opening pressure, elevated nucleated cells, elevated protein and normal glucose. CSF cytology, gram stains and cultures were unremarkable. Possibility of sarcoidosis, mycobacterium and fungal etiologies were ruled out. Other work up included but not limited to MRA head, MRV head, diagnostic cerebral angiogram, extensive serum and CSF autoimmune labs were also unremarkable. She underwent a brain biopsy which showed necrotizing granulomatous inflammation with associated dystrophic calcification. Patient was started on high dose methylprednisolone followed by a 6-week prednisone taper. There was a complete resolution of her symptoms, as well as improvement in her follow up MRI brain over a six month period.

Design/Methods

N/A.

Results

N/A.

Conclusions

We describe what we believe would be the third reported case of necrotizing granulomatous meningitis of an unknown cause in humans. This case demonstrates the response of necrotizing granulomatous meningoencephalitis in humans to steroid therapy. Yet, further studies are necessary to determine symptomatology and pathogenesis, as well as the treatment in humans.

Disclosure: Dr. Lateef has nothing to disclose. Dr. Gharaibeh has nothing to disclose. Ms. Zamir has nothing to disclose. Dr. Sheikh has nothing to disclose.

Ischemic Stroke in Neurosarcoidosis: A Retrospective Cohort Analysis

Spencer Hutto, Kevin Kyle, Denis Balaban, Nagagopal Venna

Objective

To provide a detailed analysis of the clinical features and course of ischemic stroke secondary to sarcoidosis of the CNS and to observe the effects of immunosuppression in the prevention of recurrent cerebrovascular disease.

Background

Cerebrovascular disease is rarely reported in neurosarcoidosis and constitutes one of its least well-described forms, though recognition for it has grown in the last decade with recent studies estimating a higher frequency of occurrence than previously known.

Design/Methods

Patients with ischemic stroke were included if the mechanism was directly attributable to sarcoidosis of the CNS. Patients were excluded if an alternative stroke etiology was of equal or higher likelihood than CNS sarcoidosis.

Results

Neurologic disease was the initial presenting manifestation of sarcoidosis in 8/11 (72.7%), and ischemic stroke was an inaugural manifestation of sarcoidosis in 4/11 (36.4%). Small vessel disease was the predominant ischemia subtype (10/11, 90.9%) with pontine perforating vessels (6/11, 54.5%) and lenticulostriate arteries (3/11, 27.3%) being the vessels most often affected. Vessels with a more rostral supratentorial distribution were uncommonly affected. Common neuroinflammatory accompaniments included leptomeningitis (10/11, 90.9%) and cranial nerve disease (4/11, 36.4%). Recurrent strokes occurred in 8/11 (72.7%), and recurrent neuroinflammation occurred in 7/11 (63.6%). Antiplatelet drugs were used in 6/11 (54.5%). Most patients (10/11,

90.9%) required at least two lines of immunosuppression to achieve inflammatory disease remission in this context; infliximab was the most successfully employed immunosuppressant (7/8 treatment courses, 87.5%). The presenting median modified Rankin Scale score of 4.0 improved to 2.0 over a median period of follow-up of 52.0 months.

Conclusions

Ischemic strokes in neurosarcoidosis occur in a caudal-to-rostral distribution, tend to affect small caliber blood vessels that lack collateral blood flow, and typically associate with inflammatory leptomeningeal disease.

Disclosure: Dr. Hutto has nothing to disclose. Dr. Kyle has nothing to disclose. The institution of Dr. Balaban has received research support from Biogen. Dr. Venna has nothing to disclose.

Pachymeningitis in Biopsy-Proven Sarcoidosis: Clinical Course, Radiographic Findings, Response to Treatment, and Long-Term Outcomes

Pressley Chakales, Max Herman, Ling Chen Chien, Spencer Hutto

Objective

To study the clinicoradiographic features of pachymeningeal involvement in neurosarcoidosis and its evolution over time in response to treatment.

Background

Meningeal inflammation is one of the most common forms of neurosarcoidosis, occurring in 16-69% of affected patients. While the clinical and radiographic features of leptomeningitis in neurosarcoidosis are well known, those of pachymeningitis are far less clear.

Design/Methods

Patients with a diagnosis of neurosarcoidosis seen at Emory University [01/2011-8/2021] were included if pachymeningeal involvement was evident by MRI and the patient's sarcoidosis was pathologically confirmed (from a neural or extraneural site).

Results

26/215 (12.1%) patients with neurosarcoidosis qualified for inclusion. Pathological confirmation came from neural tissue in 50%. Median age of onset was 43.5 years; most were male (16/26, 61.5%). Symptoms were primarily related to pachymeningitis in 20/26 (76.9%). Headache (19/26, 73.1%), visual dysfunction (12/26, 46.2%), and seizures (7/26, 26.9%) were the most common symptoms. All patients had cranial pachymeningitis; only a single patient undergoing spinal imaging (1/11, 9.1%) had spinal pachymeningitis. The falx cerebri (16/26, 61.5%) was the most commonly affected dural structure, but the anterior and middle cranial fossae and tentorium were frequently involved (12/26 each, 46.2%). The pachymeningeal lesions were unifocal (11/26, 42.3%) or multifocal (15/26, 57.7%) in distribution, nodular morphologically (23/25, 92.0%), and homogeneously enhancing (24/25, 96.0%). Symptomatic improvement occurred with steroids initially in 22/25 (88.0%). Ultimately, 23/26 (88.5%) required initiation of steroid-sparing immunosuppressants, including 8/26 (30.8%) eventually undergoing TNF inhibition. Pachymeningeal relapses occurred in 7/26 (26.9%). Median clinical follow-up was 48 months. Median mRS at last follow-up improved to 1.0 from 2.0 at presentation.

Conclusions

Sarcoid pachymeningitis often presents with headaches and visual dysfunction, usually affects the falx cerebri and anterior and middle cranial fossae, and tends to require steroid-sparing immunosuppressants. It has the potential to relapse, but the prospect for recovery is excellent.

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