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

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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.

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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.

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

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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.

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