Focal idiopathic eosinophilic meningoencephalitis: A case report of a novel entity

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Abstract

Common causes of eosinophilic meningitis include infectious diseases, especially parasitic infections of the central nervous system and noninfectious etiologies such as lymphoma and drug induced meningitis. However eosinophilic meningitis as a primary etiology has not been reported in the literature. We report the case of a 46-year-old old woman who presented with first time seizure. Magnetic Resonance Imaging of the brain was significant for a heterogeneous lesion involving the frontal cortex with overlying dural thickening and enhancement. Biopsy of the meninges and underlying brain parenchyma suggested a diagnosis of primary eosinophilic meningoencephalitis with no evidence of microbial involvement, vasculitis or neoplastic processes. We suggest that focal idiopathic eosinophilic meningoencephalitis may be a novel entity that responds dramatically to steroids.

Introduction

Eosinophilic meningitis is defined as meningeal inflammation in the presence of 10 eosinophils/mm³ in the cerebrospinal fluid (CSF) or eosinophils accounting for more than 10% of CSF leukocytes [1]. Parasitic infections, malignancy, medications, recreational drugs and vasculitis constitute common causes of eosinophilic meningitis and meningoencephalitis (REF). We report a middle-aged lady with eosinophilic meningoencephalitis, who despite extensive laboratory, imaging and pathological investigations did not demonstrate an underlying etiological process-leading us to suggest the term focal idiopathic eosinophilic meningoencephalitis.

Case Presentation

A 46-year-old Caucasian lady with no significant past medical history presented to the emergency department following a two-minute generalized tonic-clonic convulsion. She was in her usual state of health and had no neurological complaints prior to the seizure. There was no history of trauma, recent travel, medication use or serious infections in the past. She did not have any recent acute illnesses and there were no sick contacts of note.

Shortly after the seizure, she was back to her baseline and general physical examination revealed no major abnormalities except for laceration of her tongue. Her mental status and cranial nerve examination were intact. Motor strength, tone, bulk and sensory examination revealed no aberrations. Her coordination and gait were also normal. Hyperreflexia of the left patella and ankle were noted.

Laboratory Investigations and Imaging

Complete blood count, electrolytes, renal and liver function tests were within normal limits. Of note, there was no peripheral eosinophilia. Cerebrospinal fluid (CSF) analysis showed normal chemistry and no pleocytosis. Studies performed on the CSF for bacterial, viral, fungal, ova and parasites, tuberculosis, cryptococcal antigen and angiotensin converting enzyme (ACE) levels to check for sarcoidosis were negative. Syphilis, Human Immune deficiency virus (HIV), toxoplasma and immunological workup in the blood including anti-nuclear antigen (ANA), extractable nuclear antigens (ENA), antineutrophil cytoplasmic antibody (ANCA) and anti-double -stranded DNA antibody (Anti ds DNA) were negative. Computed tomography (CT) of the head and vessel imaging of the brain including computed tomography angiography and venogram (CTA and CTV) were normal. Electroencephalogram done shortly after the convulsion showed generalized slowing but no epileptiform discharges.

Magnetic resonance imaging (MRI) of the brain showed a single intra-axial heterogeneous lesion involving the cortex and sub cortical white matter in the anterior aspect of the right frontal lobe with cortical enhancement and perilesional edema (Figure 1). There was thickening and enhancement of the overlying dura mater as well. Diffusion weighted images (DWI) images showed diffusion restriction in the corresponding areas suggestive of vasogenic edema. Repeat MRI of the brain, performed one week later, showed interval progression of the vasogenic edema associated with gyral thickening. It also showed persistent abnormal contrast enhancement in the same region with additional development of microhemorrhages, suggestive of localized inflammatory/infectious cerebritis.

Biopsy of the affected brain parenchyma and overlying meninges revealed eosinophilic meningitis without evidence of microbial involvement. Additional biopsy findings were liquefactive necrosis of the cortex with residual “red” neurons and florid microvasculature proliferation (Figure 2A). Arteries...
and veins were surrounded by a loose infiltrate of small mature lymphocytes and eosinophils with sparse patchy extension of the inflammation along blood vessels into the necrotic cortex (Figure 2B). The overall picture thus appeared to be that of eosinophilic meningitis associated with ischemic infarction of the overlying cortex.

Our patient was treated with five days of intravenous steroids. She did not have any further seizures or neurological issues and made a complete recovery after the course of steroids. Follow up neurological evaluation after 6 months was normal, and she could return to her occupation of laboratory technician, with no cognitive or motor limitations.

Discussion

Eosinophilic meningitis can potentially be the result of certain infections of the central nervous system. However, non-infectious causes have occasionally been noted. Angiostrongyliasis, gnathostomiasis and neurocysticercosis are the most common infectious etiologies. The rat lungworm Angiostrongylus cantonensis, a food-borne zoonotic parasite, has been recognized as the primary pathogen associated with most forms of human eosinophilic meningitis or eosinophilic meningoencephalitis [2]. All patients with Angiostrongylosis related eosinophilic meningoencephalitis reported eating raw or partially cooked monitor lizard (Varanus bengalensis) several days prior to their presentation [2,3]. Angiostrongylia can also be transmitted by eating infected raw or undercooked snails, poorly cleaned contaminated vegetables or other infected intermediate hosts such as freshwater prawns, crabs or frogs [2].

Malignancies, especially leukemia and lymphoma involving the brain [4-6], medications such as non-steroidal anti-inflammatory drugs (NSAIDs) and antibiotics such as cotrimoxazole [7-9] post-myelography and certain forms of vasculitis including rheumatoid arthritis [10], primary angiitis of the central nervous system [11] and eosinophilic aseptic arachnoiditis in intravenous drug abusers [12] are less common non-infectious causes of eosinophilic meningitis/cerebritis.

In the case discussed above, no organisms were identified on histological examination of the affected tissue, with routine and special stains. No tumor cells were identified in the CSF...
or pathological specimen of the brain. Although some of the leptomeningeal veins contained inflammatory cells within their walls, there was no evidence of necrotizing vasculitis, the arteries appeared normal and no granulomas were seen. The possibility that organisms were indeed present but not represented in the sample can be considered. However adjunct testing for microorganisms in blood and CSF was found to be negative. After a meticulous review of clinical and laboratory findings, imaging, and pathology of the affected tissue, a diagnosis of primary focal eosinophilic meningitis was made.

In conclusion, we would like to propose that eosinophilic meningitis can occur as a primary (idiopathic) condition, be highly localized to a small region of the cortex and overlying meninges and can be diagnosed by careful exclusion of other infectious and non-infectious causes. Response to steroids in our case was highly satisfactory and resulted in resolution of symptomatology.

References


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