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Case Report:

Primary Amoebic (*Naegleria fowleri*) Meningoencephalitis Presenting as Status Epilepticus

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Abstract:
Primary amebic meningoencephalitis (PAM) is a rare entity. Usual presenting features are fever, headache and seizures with meningeal signs and this disease carries high mortality rate. We present a case report of PAM presenting as status epilepticus.
Key Words: Primary amebic meningoencephalitis; *Naegleria fowleri*; Status epilepticus

Introduction:
N. fowleri is the only species of Naegleria genus that is pathogenic to humans. It is ubiquitous in natural fresh water lakes and ponds (especially warm water). It causes extremely rare and sporadic CNS infections. Typically, *N. fowleri* produces primary amebic meningoencephalitis (PAM), which is clinically indistinguishable from acute bacterial meningitis.

Case Report:
A 21 years female residing at district Bilaspur of Himachal Pradesh, presented with fever, headache and vomiting for 1 week and seizures for 1 day. Fever was high grade (up to 103°F), intermittent, associated with chills and rigors. Headache was diffuse dull aching to start with, present all over the head, and later on it increased in severity. She also had vomiting 7-8 episodes per day, mainly consisting of undigested food particles, non bilious, not containing any blood, mucus. There was also history of 7-8 episodes of generalised tonic clonic seizures without regaining consciousness. General physical examination was unremarkable. Neurological examination revealed unconscious state with neck rigidity and positive Kernig's sign. The rest of the systemic examination was normal. Complete haemogram, LFT's , RFT's, and blood culture were normal. CSF examination revealed proteins 55 mg%, sugar 30 mg%, cytology 7 WBCs, mostly lymphocytes. Wet mount specimen of CSF revealed motile flagellate forms of *Naegleria fowleri*. CSF was repeated twice which revealed similar form on microscopy. MRI brain was done which was normal. Retrospectively history revealed that she had taken a bath at a local pond. On the day 1, patient was initially started empirically on ceftriaxone, vancomycin, acyclovir and steroid on the lines of acute bacterial meningitis and acute viral encephalitis and also patient was dilantinised for status epilepticus. On day 2 after CSF report, patient was started on injection amphotericin. Despite this, patient deteriorated and she expired on 5th day of admission.

Discussion:
Primary amebic meningoencephalitis is caused by free living amebas of genera *Naegleria fowleri*, Acanthamoeba and Balamuthia mandrillaris formerly known as leptomyxid amoeba. These are free living, amephizoic and opportunistic protozoa that are ubiquitous in nature. These amebas are found in soil, water, air samples from all over world. Man gets infected from inhalation of trophozoites or cysts on exposure to polluted water in ponds, swimming pools and manmade lakes. *Nageleria fowleri* is a thermophilic amoeba that grows well in tropical and subtropical climates and raised temperatures during hot summer months or warm water from power plants facilitates its growth. Human infection involves brain, lung, skin and eyes and has increased significantly since last 10 years. The portal of entry to CNS is through olfactory neuroepithelium. CNS infection occurs in two forms:

a) Primary amebic meningoencephalitis, (PAME) caused by *Naegleria fowleri*

b) Granulomatous amebic encephalitis, (GAE) caused by species of Acanthamoeba and *Balamuthia mandrillaris*.

Incubation period for *N. fowleri* is 3 to 7 days and that for acanthamoeba is unknown but perhaps more than 10 days. PAME produced by *N. fowleri* is characterized by acute fulminant meningoencephalitis with severe headache, high grade fever, nausea, vomiting, altered consciousness with signs of meningeal irritation. Cranial nerve palsies, seizures, confusion, agitation, and coma may follow and death occurs within a week if not treated early. Most patients die, usually by the seventh day.[2-5]

Recognition of primary amoebic meningoencephalitis depends on clinical suspicion based on patient history. A spinal tap usually reveals an elevated white blood cell count (>100 cells/mm3) with a high percentage of polymorphonuclear leukocytes (>70%), a depressed glucose level (<50 mg/dL), and an elevated protein level (>60 mg/dL), mimicking those of bacterial meningitis. Occasionally, amebae can be observed on Gram-stained smears. In CSF, both amoeboid and flagellate forms have been reported. If primary amoebic meningoencephalitis is suspected, a fresh-centrifuged specimen of CSF should be inspected by wet-mount preparation and fixation and staining. Special culture media are required for culture of amoeba. Serological tests performed are indirect Immunofluorescence assay (IIA), dot immunobinding assay (DIBA) and enzyme linked immunotransfer blot technique (EITB). Neuroimaging findings nonspecific brain edema suggestive of meningitis, favouring *N. fowleri,* and punctuate enhancing lesions due to acanthamoeba.[6-8]

A few patients have been successfully treated using a combination of miconazole, oral rifampin and high dose of injection amphotericin.[1,9] The risk of infection from water containing *N. fowleri* is unknown but probably small, since thousands of people swim in lakes, ponds and hot springs known to contain these organisms, and yet cases of PAM are extremely rare. This case is peculiar because we found flagellate form in CSF in contrast to amoeboid form reported in other case reports although both are known to cause PAM. PAM is known to cause seizures, but presentation as status epilepticus is rare. To the nest of our knowledge, no case of PAM has been reported from the Sub-Himalayan region. PAM should be suspected in patients with acute meningitis who do not respond to conventional treatment and who have fulminant course.

References:

1. Brown RL. Successful treatment of primary amebic meningoencephalitis (PAM). *Arch Intern Med* 1991;151:1201-1202
2. Jain R, Prabhakar S, Modi M, Bhatia R, Sehgal R. *Naegleria* meningitis: a rare survival. *Neurol India* 2002;50:470-472.
3. Martinez AJ. Free living amoebas: Infection of the central nervous system. *Mt Sinai J Med* 1993;60:271-278.
4. Shamsizadeh A, Zaker N, Maraghi S, Momen A. A Primary Amebic Meningoencephalitis. *Pak J Med Sci* 2006;22:471-473.
5. Tungikar SL , Kulkarni AG, Deshpande AD, Gosavi VS. Primary Amebic Meningoencephalitis. *JAPI* 2006;54:327-329.
6. Rodriguez R, Mendez O, Molina O, Luzardo G. CNS infection by free living amebasc Report of 3 Venezuelan cases. *J Rev Neurol* 1998;26:1005-1008.
7. Garcia TJ, Sotolongo GF, Cepero NF, Ibarra SE, Garcia OJ. Primary amebic meningoencephalitis. Report of suspicious case and review of the medical literature. *Rev Cubana Med Trop.* Sep-Dec 1978;30(3):161-166.
8. Jain R, Prabhakar S, Modi M, Bhatia R, Sehgal R. Naegleria meningitis: a rare Survival. *Neurol India* 2002;50:470-472.
9. Singh SN, Patwari AK, Dutta R, Taneja N, Anand VK. Naegleria meningitis. *Ind Pediatr* 1998;35:1012-1015.