CLINICAL CASE REPORT

A local case of fulminant primary amoebic meningoencephalitis due to *Naegleria fowleri*

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ABSTRACT:
Primary amoebic meningoencephalitis is an extremely rare, predominantly fulminant central nervous system infection caused by the amoeba *Naegleria fowleri*, first described in Australia in 1965. Despite the ubiquitous presence of *N. fowleri*, as few as 300 cases of infection have since been reported worldwide, with a case fatality rate approaching 98%. A combination of low index of suspicion, non-specific clinical findings and largely ineffective treatment modalities make this rapidly progressive meningencephalitis virtually impossible to treat. Early and aggressive treatment utilising intravenous and intrathecal routes by a multidisciplinary team of neurosurgeons, intensivists and microbiologists is required. Presented is a case of a 56-year-old man who presented to the Gold Coast University Hospital in Queensland, Australia, with rapidly progressive primary amoebic meningoencephalitis. He received maximal therapy and died of his disease while in hospital.

Keywords:
amoeba, Australia, *Naegleria fowleri*, meningencephalitis, pyogenic.

FULL ARTICLE:
A man aged 56 years, recently returned from work in Central Queensland, presented to the emergency department of the Gold Coast University Hospital in Queensland, Australia, complaining of 36 hours of severe headache, photophobia, nausea, vomiting and neck stiffness. On examination, his Glasgow Coma Scale score was 14 (eye opening 3, verbal response 5, motor response 6), he was febrile (38.5°C), photophobic, and neck stiffness was objectively evident. Cerebrospinal fluid obtained by lumbar puncture was turbid, with glucose <0.3 mmol/L (reference 2.8–4.0 mmol/L), protein 6700 mg/L (reference 150–500 mg/L), white blood cell count 1460 × 10^6/L (90% polymorphs) and red blood cell count 460 × 10^6/L.

The patient was empirically started on ceftriaxone, benzylpenicillin and dexamethasone. Over the subsequent 2 hours a decline in Glasgow Coma Scale score to 10 (eye opening 2, verbal response 2, motor response 6) necessitated admission to the intensive care unit (ICU). Initial brain CT was normal.

Wet mount microscopy identified free-living amoeba, and diagnosis of primary amoebic meningoencephalitis was made within hours of presentation to the emergency department. Multiplex polymerase chain reaction identified *Naegleria* species. The patient was electively taken to theatre for insertion of a ventriculostomy drain to allow intrathecal administration of amphotericin. Treatment consisted of intrathecal amphotericin 1.5 mg daily, amphotericin 50 mg intravenously (IV) every 12 hours, rifampicin 600 mg IV daily, azithromycin 500 mg IV daily, fluconazole 800 mg IV daily, dexamethasone 8 mg twice daily and chlorpromazine 50 mg IV every 4 hours.

On day 2 after ICU admission, the patient demonstrated profound and maintained sympathetic nervous system activation, with heart rate >150 beats per minute, systolic blood pressure >180mmHg and respiratory rate >40 breaths per minute. Brain CT results remained unchanged from admission images, echocardiography showed a vigorous heart, electroencephalogram ruled out status epilepticus and his vital signs showed no response to fluid/electrolyte replacement, adenosine, analgesia or sedation. Despite early aggressive management the patient had bilateral fixed and dilated pupils on day 3 of his ICU admission. Further CT imaging demonstrated extensive oedema in the frontal, temporal and occipital lobes, global sucal effacement and posterior fossa mass effect. After discussion with the family regarding clinical progression of disease, the decision was made to withdraw treatment and provide palliation, and the patient died within 72 h of presentation.

**Discussion**

*Naegleria fowleri* is a free-living amoeba typically inhabiting warm bodies of fresh water and soil. It was first linked as the causative organism to PAM in 1965 by Australian pathologists Fowler and Carter. Epidemiological studies reveal PAM typically affects young males exposed to warm bodies of fresh water. A review of 111 cases of PAM occurring in the USA between 1962 and 2008 found the average length of time from exposure to onset of symptoms for these cases was 5 days, the average length of time from onset of symptoms until death was 5.3 days and average length of time from exposure to death was 9.9 days.

Upon exposure to contaminated water, a person is infected by migration of *N. fowleri* across the nasal mucosa and via the criiform plate to the olfactory bulbs. From there the organism gains access to the brain parenchyma, causing a necrotising meningoencephalitis. The patient in the present study had a history of significant exposure to untreated fresh water. He had recently performed physical work on a property in Central Queensland and had swum, on two separate occasions, in stagnant water that was also accessed by cattle. According to his family, he had a habit of irrigating his nostrils in the shower after work, which he had most recently done in the days preceding his presentation to hospital. Water used to irrigate his nose was untreated bore water.

Effective treatment remains elusive and the case fatality rate approaches 98%. The mainstay of treatment has been amphotericin B, intravenously and sometimes intrathecally. However, recent in vitro and in vivo studies of amphotericin B, miltefosine and chlorpromazine in *N. fowleri*-infected mice demonstrated greater survival in those treated with chlorpromazine than with amphotericin (75% v 40%). The optimal route of delivery of chlorpromazine is not known.
In the case reports of those extremely rare patients who have been thought to be PAM survivors it has been postulated that Acanthamoeba species or Balamuthiamandrillaris may have been the causative organism. These typically produce a more subacute course. The few patients reported to have survived proven PAM have been young children presenting within a few hours of symptom onset, allowing aggressive early management.

**Conclusion**

Due to its rare and rapidly progressive nature, and the need for a high index of suspicion, PAM will likely continue to result in high mortality. A high index of suspicion and early involvement of infectious disease specialists and microbiologists are required so that rapid, aggressive intravenous and intrathecal therapy are possible.

**REFERENCES:**


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