Another Killer of the Australian Bush: A Rapidly Fatal Meningoencephalitis in a Child

(See pages 1375–1376 for the Photo Quiz.)

Figure 1. Magnetic resonance imaging of the brain showing diffuse cortical increased signal on T2-weighted imaging.

Diagnosis: *Naegleria fowleri* primary amoebic meningoencephalitis.

The magnetic resonance imaging of the brain showed diffuse cortical increased signal on T2-weighted imaging (Figure 1). There was hemorrhagic meningitis in all areas of the brain surface. Cerebellar tonsil herniation with marked raised intracranial pressure subsequently developed.

Phase contrast microscopy of an unstained cerebrospinal fluid (CSF) specimen showed numerous motile amoeba (Figure 2). A Wright’s stain demonstrated multivacuolated trophozoites 15–25 μm in diameter (Figure 3). These were confirmed to be *N. fowleri* by nucleic acid amplification.

A sibling of the patient had died of a meningitic illness 8 years before the patient presented to our institution. No definitive cause was apparent for that death.

Primary amoebic encephalitis is caused by *N. fowleri* and usually occurs in previously healthy children who have had contact with untreated fresh water. This would include natural bodies of water and domestic sources of water, such as pools, wells, and—as in this case—bores [1–3]. In particular, there have been 2 reported cases from Arizona that occurred in children who had been exposed to well water [3]. Access to the central nervous system is believed to occur by direct invasion through the nasal mucosa and the cribriform plate. The onset of symptoms occurs 2–5 days after the last contact with fresh water. There is a rapid onset of fever, headache, meningismus,
and altered mental state with seizures. This usually results in a rapidly fatal meningoencephalitis. In contrast, granulomatous amoebic encephalitis is caused by *Acanthamoeba* species, *Balamuthia mandrillaris*, or *Sappinia diploidea* and is an opportunistic chronic infection that usually occurs in immunocompromised individuals [4].

The initial cases of *N. fowleri* meningoencephalitis were first described by Fowler and Carter [5] in southern Australia in 1965, but it has since been described worldwide [1, 2, 4, 6, 7]. The reported mortality can exceed 95% [8]. Pathogenic *N. fowleri* multiply in fresh water, and growth can occur within a temperature range of 13°C–46°C, although most have been reported in water temperatures of 25°C–42°C. The presence of *N. fowleri* in fresh water is directly related to water temperature [3, 9].

The diagnosis of primary amoebic meningoencephalitis can be difficult if a wet unstained preparation of CSF is not examined for the motile trophozoites. Molecular diagnostic tests are not generally easily available but are sensitive and specific [10]. Amphotericin B is the treatment of choice and can be

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**Figure 2.** Phase contrast microscopy of a cerebrospinal fluid specimen from the patient (original magnification, ×400). 1, lymphocyte; 2, motile *Naegleria fowleri* trophozoites; 3, erythrocyte.

**Figure 3.** Wright’s stain of a cerebrospinal fluid specimen from the patient (original magnification, ×1000). Clumped *Naegleria fowleri* trophozoites each showing a single large nucleus with a dense karyosome (black arrow) and prominent cytoplasmic vacuolation (white arrow) are shown. A lymphocyte and red blood cell are also shown for size comparison.
given both intravenously and intrathecally. A variety of combination therapies have been tried, including fluconazole, rifampicin, chloramphenicol, and azithromycin [4, 11, 12]. Nevertheless, associated mortality remains high.

This patient was treated with intravenous and intraventricular amphotericin, intravenous rifampicin, and azithromycin. Despite aggressive and appropriate therapy in the intensive care unit, her condition rapidly deteriorated, and she died within 72 h after presentation. A recommendation was made to the family to install an in-line bacterial filter for all household water originating from the bore to prevent future episodes of amebic meningoencephalitis.

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Robert Norton,1,2 Patrick Harris,1,2 Pat Ryan,3 and Scott Simpson4
1Pathology Queensland and 2Townsville Hospital, Townsville, Queensland, Australia

References