

DISEASES/CONDITIONS

Amebic Encephalitis (*Naegleria fowleri*, *Balamuthia mandrillaris*, *Acanthamoeba*)

Merlin reporting code = 13620

Case report form (CRF): [Primary Amebic Meningoencephalitis CRF](#)
PAPER CRF REQUIRED

Naegleria fowleri Causing Primary Amebic Meningoencephalitis (PAM)

Clinical description

N. fowleri is a free-living ameboflagellate that invades the brain and meninges via the nasal mucosa and olfactory nerve to cause acute, fulminant hemorrhagic meningoencephalitis (primary amebic meningoencephalitis – PAM), primarily in healthy children and young adults with a recent history of exposure to warm fresh water. Initial signs and symptoms of PAM begin 1 to 14 days after infection and include sudden onset of headache, fever, nausea, vomiting, and stiff neck accompanied by positive Kernig's and Brudzinski's signs. In some cases, abnormalities in taste or smell, nasal obstruction and nasal discharge may be seen. Other symptoms may include photophobia, mental-state abnormalities, lethargy, dizziness, loss of balance, other visual disturbances, hallucinations, delirium, seizures, and coma. After the onset of symptoms, the disease progresses rapidly and usually results in death within 3 to 7 days. Although a variety of treatments have been shown to be active against amebae *in vitro* and have been used to treat infected persons, most infections have still been fatal.

Laboratory criteria for case classification

Detection of *N. fowleri*

- Organisms in CSF, biopsy, or tissue specimens;
- OR
- Nucleic acid (e.g., polymerase chain reaction) in CSF, biopsy, or tissue specimens;
- OR
- Antigen (e.g., direct fluorescent antibody) in CSF, biopsy, or tissue specimens.

Case classification

Confirmed: A clinically compatible illness in a person with laboratory evidence. When available, molecular characterization should be reported (e.g. genotype).

Comments

N. fowleri may cause clinical illness similar to bacterial meningitis, particularly in its early stages. Definitive diagnosis by a reference laboratory may be required. Unlike *B. mandrillaris* and *Acanthamoeba* spp., *N. fowleri* is commonly found in CSF.

Balamuthia mandrillaris Disease

Clinical description

B. mandrillaris is an opportunistic free-living amoeba that may invade the brain through the blood, probably from a primary infection in the skin (from ulcers or dermatitis), the sinuses and middle ear (from rhinitis, sinusitis, or otitis media), or via organ transplantation. The incubation period is not well characterized but has been observed to range from 2 weeks to months or possibly years. Once in the brain, the amoebae can cause meningoencephalitis or granulomatous amebic encephalitis (GAE). The

amebae may also invade the brain via the nasal mucosa and olfactory nerve. *B. mandrillaris* GAE often has a slow and insidious onset and develops as a subacute or chronic disease lasting several weeks to months; however, *B. mandrillaris* infections associated with organ transplantation have an especially rapid clinical course. *B. mandrillaris* GAE generally affects persons who are immunosuppressed from a variety of causes (e.g., HIV/AIDS, IV drug use). However, cases have also occurred in young children and older adults with no obvious signs of immunosuppression. In some instances, affected individuals have had a relatively rapid clinical course. Initial symptoms of *B. mandrillaris* GAE may include headache, photophobia, and stiff neck accompanied by positive Kernig's and Brudzinski's signs. Other symptoms may include nausea, vomiting, low-grade fever, muscle aches, weight loss, mental-state abnormalities, lethargy, dizziness, loss of balance, cranial nerve palsies, other visual disturbances, hemiparesis, seizures, and coma. Painless skin lesions appearing as plaques a few millimeters thick and one to several centimeters wide have been observed in some patients, especially patients outside the U.S., preceding the onset of neurological symptoms by 1 month to approximately 2 years. Once the disease progresses to the acute stage, it is generally fatal within weeks or months. However, a few patients have survived this infection.

Laboratory criteria for case classification

Detection of *B. mandrillaris*

- Organisms in CSF, biopsy, or tissue specimens;
- OR
- Nucleic acid (e.g., polymerase chain reaction) in CSF, biopsy, or tissue specimens;
- OR
- Antigen (e.g., direct fluorescent antibody) in CSF, biopsy, or tissue specimens.

Case classification

Confirmed: A clinically compatible illness in a person with laboratory evidence. When available, molecular characterization should be reported (e.g. genotype).

Comments

B. mandrillaris and *Acanthamoeba* spp. may cause clinically similar illnesses and may be difficult to differentiate using commonly available laboratory procedures. Definitive diagnosis by a reference laboratory may be required. A negative test on CSF does not rule out *B. mandrillaris* infection because the organism load in the CSF is often low.

Acanthamoeba Disease (Excluding Keratitis)

Clinical description

The genus *Acanthamoeba* includes several species of opportunistic free-living amebae that may invade the brain through the blood, probably from a primary infection in the skin (from ulcers or dermatitis) or the sinuses and middle ear (from rhinitis, sinusitis, or otitis media). Once in the brain, the amebae cause a granulomatous amebic encephalitis (GAE). The amebae may also invade the brain via the nasal mucosa and olfactory nerve. *Acanthamoeba* GAE has a slow and insidious onset and develops as a subacute or chronic disease lasting several weeks to months. *Acanthamoeba* GAE generally affects persons who are immunosuppressed from a variety of causes (e.g., HIV/AIDS, diabetes, organ transplantation). However, a few cases have been described in individuals with no obvious signs of immunosuppression. Initial symptoms of *Acanthamoeba* GAE may include headache, photophobia, and stiff neck accompanied by positive Kernig's and Brudzinski's signs. Other symptoms may include nausea, vomiting, low-grade fever, muscle aches, weight loss, mental-state abnormalities, lethargy, dizziness, loss of balance, cranial nerve palsies, other visual disturbances, hemiparesis, seizures, and

coma. Once the disease progresses to the acute stage, it is generally fatal within weeks or months. However, a few patients have survived this infection.

Laboratory criteria for case classification

Detection of *Acanthamoeba* spp.

- Organisms in CSF, biopsy, or tissue specimens;
- OR
- Nucleic acid (e.g., polymerase chain reaction) in CSF, biopsy, or tissue specimens;
- OR
- Antigen (e.g., direct fluorescent antibody) in CSF, biopsy, or tissue specimens.

Case classification

Confirmed: A clinically compatible illness in a person with laboratory evidence. When available, species designation and molecular characterization (e.g., genotype) should be reported.

Comments

Acanthamoeba and *B. mandrillaris* may cause similar clinical illnesses and may be difficult to differentiate using commonly available laboratory procedures. Definitive diagnosis by a reference laboratory may be required. Several species of *Acanthamoeba* are associated with infection (i.e., *A. castellanii*, *A. culbertsoni*, *A. hatchetti*, *A. healyi*, *A. polyphaga*, *A. rhyodes*, *A. astonyxis*, *A. lenticulata* and *A. divionensis*). A negative test on CSF does not rule out *Acanthamoeba* infection because the organism is not commonly present in CSF.

Acanthamoeba Keratitis

Clinical description

Acanthamoeba keratitis is a local infection of the cornea (outer layer of the visual pathway of the eye) caused by a microscopic, free-living amoeba belonging to the genus *Acanthamoeba*. Symptoms include foreign body sensation, photophobia, decreased visual acuity, tearing, pain, and redness of the eye. It occurs most typically among health, contact lens users, but can occur in anyone. Although treatable with topical medications, affected individuals are at risk for permanent visual impairment or blindness. *Acanthamoeba* organisms are ubiquitous in nature and can be found in bodies of water (e.g., lakes and oceans), soil, and air.

Laboratory criteria for case classification

Confirmatory:

Detection of *Acanthamoeba* spp.

- Organisms in corneal scraping or biopsy specimens;
- OR
- Nucleic acid (e.g., polymerase chain reaction) in corneal scraping or biopsy specimens;
- OR
- Antigen (e.g., direct fluorescent antibody) in corneal scraping, or biopsy specimens.

Presumptive:

Identification of *Acanthamoeba* trophozoites or cysts using confocal microscopy

Case classification

Confirmed: A clinically compatible illness in a person with confirmatory laboratory evidence. When available, species designation and molecular characterization (e.g., genotype) should be reported.

Probable: A clinically compatible illness in a person with presumptive laboratory evidence.

[Return to Table of Contents](#)