Diseases and Conditions

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

Merlin disease code=13629 Amebic Encephalitis (*Naegleria fowleri*)
Merlin disease code=13625 Amebic Encephalitis (*Balamuthia mandrillaris*)
Merlin disease code=13621 Amebic Encephalitis (*Acanthamoeba*)
Case report form (CRF): *Primary Amebic Meningoencephalitis CRF*
PAPER CRF REQUIRED

*Naegleria fowleri* Causing Primary Amebic Meningoencephalitis (PAM)

Clinical criteria for case classification
An infection presenting as meningoencephalitis or encephalitis. The clinical presentation of PAM is like that of acute meningitis caused by other pathogens and symptoms include headache, nausea, vomiting, anorexia, fever, lethargy, and stiff neck. Disorientation, mental status changes, seizure activity, loss of consciousness, and ataxia may occur within hours of initial presentation.

Laboratory criteria for case classification
Confirmatory:
Detection of *N. fowleri* antigen or nucleic acid from a clinical specimen (e.g., direct fluorescent antibody, polymerase chain reaction, immunohistochemistry).

Presumptive:
Either of the following:
- Visualization of motile amebae in a wet mount of cerebrospinal fluid (CSF) or
- Culture of *N. fowleri* from a clinical specimen.

Epidemiological criteria for case classification
Not applicable.

Case classification
Confirmed:
A clinically compatible illness in a person with confirmatory laboratory evidence.

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

Criteria to distinguish a new case from previous reports
Not applicable.

Comments
*N. fowleri* might cause clinically similar illness to bacterial meningitis, particularly in its early stages. Definitive diagnosis by a reference laboratory is required. Unlike *Balamuthia mandrillaris* and *Acanthamoeba* species, *N. fowleri* is commonly found in the CSF of patients with PAM. After the onset of symptoms, the disease progresses rapidly and usually results in death within 3 to 7 days. Patients presenting with the above clinical criteria and found to have a history of recreational freshwater exposure in the two weeks prior to presentation or are known to have performed nasal irrigation (e.g., use of a neti pot for treatment of sinus conditions or practice ritual ablution including nasal rinsing) in the absence of another explanation for their condition should be investigated further. Urgent confirmatory testing and treatment should be initiated.
**Balamuthia mandrillaris Disease**

**Clinical criteria for case classification**
An infection presenting as meningoencephalitis or encephalitis, disseminated disease (affecting multiple organ systems), or cutaneous disease. Granulomatous amebic encephalitis (GAE) can include general symptoms and signs of encephalitis such as early personality and behavioral changes, depressed mental status, fever, photophobia, seizures, nonspecific cranial nerve dysfunction, and visual loss. 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 neurologic symptoms by 1 month to approximately 2 years.

**Laboratory criteria for case classification**

**Confirmatory:**
Detection of *B. mandrillaris* antigen or nucleic acid or nucleic acid (e.g., PCR, immunohistochemistry) from a clinical specimen (e.g., tissue).

**Supportive:**
Culture of *B. mandrillaris* from a clinical specimen (e.g., tissue).

**Epidemiological criteria for case classification**
Not applicable.

**Case classification**

**Confirmed:**
A clinically compatible illness in a person with confirmatory laboratory evidence.

**Suspect:**
A clinically compatible illness in a person with supportive laboratory evidence.

**Criteria to distinguish a new case from previous reports**
Not applicable.

**Comments**
*B. mandrillaris* and Acanthamoeba species can cause clinically similar illnesses and might be difficult to differentiate using commonly available laboratory procedures. Definitive diagnosis by a reference laboratory is required. A negative test on CSF does not rule out *B. mandrillaris* infection because the organism is not commonly present in the CSF. Once the disease progresses to neurologic infection, it is generally fatal within weeks or months; however, a few patients have survived this infection. Patients presenting with the above clinical criteria who have received a solid organ transplant should be further investigated to determine if the infection was transmitted through the transplanted organ. An investigation of the donor should be initiated through notification of the organ procurement organization and transplant center.
Acanthamoeba Disease (Excluding Keratitis)

Clinical criteria for case classification
An infection presenting as meningoencephalitis or encephalitis, disseminated disease (affecting multiple organ systems), or cutaneous disease. Acanthamoeba species GAE presents similarly to B. mandrillaris GAE with early personality and behavioral changes, depressed mental status, fever, photophobia, seizures, nonspecific cranial nerve dysfunction, and visual loss. Skin lesions and sinus disease may also be seen.

Laboratory criteria for case classification
Confirmatory:
Detection of Acanthamoeba species antigen or nucleic acid (e.g., PCR, immunohistochemistry) from a clinical specimen (e.g., tissue).

Supportive:
Culture of Acanthamoeba species from a clinical specimen (e.g., tissue).

Epidemiological criteria for case classification
Not applicable.

Case classification
Confirmed:
A clinically compatible illness in a person with confirmatory laboratory evidence.

Suspect:
A clinically compatible illness in a person with supportive laboratory evidence.

Criteria to distinguish a new case from previous reports
Not applicable.

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