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Free-living Amebae Infections  
2012 Case Definition  
Free-living Amebae Infections  
2012 Case Definition  
NOTE:  
A surveillance case definition is a set of uniform criteria used to define a disease for public health surveillance. Surveillance case definitions enable public health officials to classify and count cases consistently across reporting jurisdictions. Surveillance case definitions are not intended to be used by healthcare providers for making a clinical diagnosis or determining how to meet an individual patient’s health needs.  
CSTE Position Statement(s)  
11-ID-15  
Subtype(s)  
Acanthamoeba  
disease (excluding keratitis)  
Acanthamoeba  
keratitis  
Balamuthia mandrillaris  
disease  
Naegleria fowleri  
causing primary amebic meningoencephalitis (PAM)  
Subtype(s) Case Definition  
Expand All  
Acanthamoeba  
disease (excluding keratitis)  
Clinical Description  
The genus  
Acanthamoeba  
includes several species of opportunistic free-living amebae that might invade the brain through the blood, probably from a primary infection in the skin (from ulcers or dermatitis) or sinuses. Once in the brain, the amebae cause granulomatous amebic encephalitis (GAE).  
Acanthamoeba  
GAE has a slow and insidious onset and develops into a subacute or chronic disease lasting several weeks to months.  
Acanthamoeba  
GAE affects both immunocompetent persons and persons who are immunosuppressed from a variety of causes (e.g., HIV/AIDS, organ transplantation). Initial symptoms of  
Acanthamoeba  
GAE might include headache, photophobia, and stiff neck accompanied by positive Kernig’s and Brudzinski’s signs. Other symptoms might 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 neurologic infection, it is generally fatal within weeks or months. However, a few patients have survived this infection.  
Laboratory Criteria For Diagnosis  
Laboratory-confirmed  
Acanthamoeba  
spp. infections (excluding keratitis) are defined as the 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 that is laboratory confirmed.\*  
Comments  
Acanthamoeba  
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  
infection because the organism is not commonly present in the 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 ameba 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 healthy, 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 Diagnosis  
Laboratory-confirmed  
Acanthamoeba  
spp. keratitis infections are defined as the 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.  
Case Classification  
Probable  
A clinically compatible illness with positive identification of  
Acanthamoeba  
trophozoites or cysts using confocal microscopy.  
Confirmed  
A clinically compatible illness that is laboratory confirmed.\*  
Balamuthia mandrillaris  
disease  
Clinical Description  
B. mandrillaris  
is an opportunistic free-living ameba that can invade the brain through the blood, probably from a primary infection in the skin (from ulcers or dermatitis), sinuses, 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 amebae can cause meningoencephalitis and/or granulomatous amebic encephalitis (GAE).  
B. mandrillaris  
GAE often has a slow, insidious onset and develops into 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 affects both immunocompetent persons and persons who are immunosuppressed from a variety of causes (e.g., HIV/AIDS, organ transplantation). Initial symptoms of  
B. mandrillaris  
GAE might include headache, photophobia, and stiff neck accompanied by positive Kernig’s and Brudzinski’s signs. Other symptoms might 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 neurologic symptoms by 1 month to approximately 2 years. Once the disease progresses to neurologic infection, it is generally fatal within weeks or months; however, a few patients have survived this infection.  
Laboratory Criteria For Diagnosis  
Laboratory-confirmed  
B. mandrillaris  
infection is defined as the 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 that is laboratory confirmed.\*  
Comments  
B. mandrillaris  
and  
Acanthamoeba  
spp. 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. A negative test on CSF does not rule out  
B. mandrillaris  
infection because the organism is not commonly present in the CSF.  
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 might be seen. Other symptoms might 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 Diagnosis  
Laboratory-confirmed  
N. fowleri  
infection is defined as the 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 that is laboratory confirmed.\*  
Comments  
N. fowleri  
might cause clinically similar illness to bacterial meningitis, particularly in its early stages. Definitive diagnosis by a reference laboratory might be required. Unlike  
Balamuthia mandrillaris  
and  
Acanthamoeba  
spp.,  
Naegleria fowleri  
is commonly found in CSF.  
Comments  
\*When available, species designation and molecular characterization (e.g., genotype) should be documented.  
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NNDSS receives and shares case data from state, local, and territorial health departments to help public health monitor, control, and prevent serious diseases.  
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