

SMALLPOX

Disease Overview

Smallpox was a systemic viral disease caused by the variola virus, a species of the Orthopoxvirus.

Naturally occurring smallpox disease was eradicated by 1977 through a worldwide vaccination program. Smallpox vaccine provides cross-protection against all orthopox viruses and is used to protect laboratory workers against these viruses. There is no active vaccination program.

Symptoms

Smallpox generally presented with a characteristic skin eruption. Prior to the appearance of the rash the early symptoms of smallpox were similar to influenza, including a sudden onset of high fever, malaise, headache, prostration, severe backache, and occasional abdominal pain and vomiting. After 2 to 4 days the fever subsided and there was a characteristic “centrifugal rash” distribution (appearing first in the oral mucosa/ palate and then progressing to the face and extremities, including the palms and soles, and later on the trunk). This deep-seated rash had individual lesions containing the infectious virus which progressed through all the phases of macules, papules, vesicles, pustules and then crusted scabs that fell off 3 to 4 weeks after the appearance of the rash. Atypical presentations include flat velvety lesions that do not evolve into pustules and more severe forms with confluent or hemorrhagic lesions. Smallpox was frequently confused with varicella (chickenpox).

There are two types of the smallpox, each with a different clinical course. Variola minor had a case fatality rate of less than 1%; Variola major had a case fatality rate among unvaccinated populations ranging from 20% to 50% or higher.

Previously vaccinated individuals did not experience a change in prodromal illness; however the smallpox rash was significantly modified to the extent that only a few highly atypical lesions occurred and maturation of lesions was accelerated with crusting by the 10th day.

Reservoir

Humans. Remaining variola (smallpox) virus stocks are kept in two World Health Organization (WHO) reference laboratories (United States and Russia).

Mode of Transmission

Smallpox was spread by respiratory tract droplets or by direct or indirect contact with the virus shed from skin lesions. Airborne spread was thought to be less likely, but transmission over significant distances has been documented. The conjunctivae or the placenta were occasional portals of entry. The virus is stable in dried form for months and has been transmitted by fomites such as bed linen.

Incubation period

The incubation period was from 7 to 19 days, typically 10 to 14 days to the onset of illness and 2 to 4 more days to the onset of the rash.

Period Communicability

Infectivity occurred at any time from the development of the rash to the disappearance of all scabs, approximately 3 weeks. Transmission was highest in the first week via droplet spread following appearance of the earliest oropharyngeal lesions and subsequent oropharyngeal excretion of virus.

Risk Factors

Smallpox research scientists.

Susceptibility among the unvaccinated is universal. Individuals who have been vaccinated in the past may have partial immunity. Canadians born in 1972 or later have not been routinely immunized against smallpox (unless immunized for travel to other countries); therefore, most are fully susceptible. Discontinuation of vaccination for travel was recommended by the WHO in 1980 and was no longer required by any country by 1982.

Surveillance Case Definition

Confirmed Case

Laboratory confirmation of infection:

- Isolation of variola virus from an appropriate clinical specimen.
- OR
- detection of variola virus nucleic acid.

Probable Case

- Clinical evidence of illness in a person who is epidemiologically linked to a laboratory confirmed case or to a probable case.
- OR
- Laboratory evidence of infection:
 - negative stain electron microscopic identification of variola virus in an appropriate clinical specimen.

Suspect Case

- Clinical evidence of illness in a person who is not epidemiologically linked to a laboratory confirmed case or to a probable case of smallpox.
- OR
- Atypical lesion known to be associated with the variola virus on a person who is epidemiologically linked to a laboratory confirmed or probable case.

Clinical Evidence

Smallpox is characterized by a febrile prodrome consisting of fever $> 38.3^{\circ}\text{C}$ and systemic symptoms (prostration, headache, back pain, abdominal pain and/or vomiting), which generally lasts one to four days and is followed by the development of a characteristic rash. The rash consists of deep, firm, well-circumscribed pustules that are mostly all in the same stage of development. The lesions are characteristically umbilicated. The lesions initially appear as macules, evolving into papules, vesicles and then pustules in a matter of days. Finally, crusted scabs form; they then fall off several weeks after the initial appearance of the rash. Lesions initially appear in the oral mucosa/palate and then progress in a centrifugal pattern to involve the face, arms, legs, palms, and soles. Atypical presentations include flat velvety lesions that do not evolve into pustules and more severe forms with confluent or hemorrhagic lesions.

Diagnosis and Laboratory Guidelines

Smallpox can be diagnosed by either isolation of the virus or by detecting its DNA using a PCR test. Appropriate clinical specimens are blood, lesion fluid and crust material. Specimen collection for the purpose of confirming a suspect case of smallpox must be done under strict isolation and preferably by a recently immunized person. Any testing related to suspected smallpox should be carried out under level 4 containment facilities at National Microbiology Laboratory in Winnipeg (NML). Transport of any containment level 4 should involve an activation of the Emergency Response Assistance Plan (ERAP) 24 hours prior to shipping. A preliminary result for either isolation or molecular detection can be expected 48 hours after reception of the sample at the NML.

Notification and Reporting

Per Policy 2.2 Disease and Event Notification to OCMOH and Disease and Event Reporting section.

- **CD Urgent Notification** for all confirmed cases.
- Routine Surveillance (RDSS) for all confirmed cases.

Case Management

Education

Investigation

Identification and confirmation of case.

Exclusion/Social Distancing

Strict isolation of case including negative pressure room.

Treatment

There is no cure or specific treatment. Administering smallpox vaccine (vaccinia vaccine) within four days after exposure can alleviate illness in nearly all cases. Once an individual shows symptoms, however, treatment is limited to supportive therapy and antibiotics to treat secondary bacterial infections.

Immunization

Vaccination of public health and health care personnel involved in the case investigation and clinical management is indicated.

Contact Management

Prevention and eradication of smallpox was dependent on isolation and vaccination. Vaccination of public health staff and health care workers, and first responders may also be indicated.

Education

Investigation

Identification and surveillance of contacts (including daily monitoring of temperature).

Exclusion/Social Distancing

Isolation of contacts that develop fever.

Prophylaxis

Immunoprophyllaxis:

Historical strategy was to immunize all contacts and those residing in the immediate vicinity (ring vaccination). Due to the relatively long incubation period for smallpox, historical data demonstrated that immunization within 2 to 3 days of exposure may protect against clinical disease, and if given within 4 to 5 days, may decrease the risk of death.

The vaccine consisted of a live Vaccinia virus, which was a "pox"-type virus related to smallpox. There were significant side effects and risks associated with this vaccine. In an outbreak situation and a risk of infection exists for an individual, there are no absolute contraindications to immunization.

Outbreak Management

Activate the local outbreak plan when an outbreak is declared. One case is considered an outbreak.

Management of Special Situations

Potential Agents of Bioterrorism

Smallpox has been identified as a potential agent for bioterrorism; however, the risk is low due to storage of the virus limited to two secure locations in the world (United States and Russia).