Hansen’s Disease (Leprosy)

By Kameron Petok

Dr. Gerhard Henrik Armauer Hansen of Norway was the first person to identify the Mycobacterium leprae (the etiologic agent of Hansen’s disease) under a microscope in 1873. This is why Hansen’s disease is also known as leprosy. While it used to be a widespread and feared phenomenon, Hansen’s disease is now much rarer and easily treated. In much earlier history, people associated it with such conclusions as being a punishment from God, a curse, and a hereditary disease. There is a sort of stigma associated with its appearance on the skin of infected individuals that makes people think it is highly contagious. But it is not. The discovery of the bacterium proved that it was caused by a germ rather than inherited or sent by sin or by God. In the time of the 20th century, universal treatment for leprosy was giving painful injections of oil found in the Chaulmoogra nut to infected individuals. Though it had efficacy in the short term, its long term effects were not efficient. Later in 1941, the drug Promin was started as a treatment for leprosy but its painful injections did not outweigh its treatment benefits. Then, in the 1950’s, dapsone pills became the main method of treatment but M. leprae developed a resistance to the drug. Finally, in the 1970’s, a multiple drug treatment approach for Hansen’s disease was underwent successful trials on the island of Malta in the Mediterranean Sea. In 1981, the World Health Organization started suggesting this multiple drug treatment which included dapsone, rifampicin, and clofazimine. It takes around six months to one or more years for this approach to work though. This is still the main method used today. Depending on the condition of the disease, regimens for doses of medicine can vary from taking the drugs monthly for a year if the leprosy is multibacillary (six or more skin lesions due to the bacteria) and taking monthly doses of rifampicin and daily doses of dapsone for a period of six months is it is paucibacillary (containing a very small number of bacilli/minor infection). This combination of drugs is the best known to prevent further nerve damage, deformity, disability due to damaged nerves, and further transmission by infected individuals. There is no current vaccine for M. leprae specifically, though some regions with high cases of Leprosy outbreak use the Bacillus Calmette Guerin (BCG) vaccine. It is used as both a Tuberculosis and Leprosy vaccine because Mycobacterium tuberculosis and Mycobacterium leprae are similar. But the effectiveness of this vaccine is very low which is illustrated by a study that showed an overall 20% cure rate for infected individuals.

Mycobacterium is the genus while the species is leprae in proper genetics terms. The representative bacillus measures between one to eight microns in length. M. leprae has an attraction for nerve cells, causing the symptoms of numbness and loss of feeling on peoples’ skin. The target for the bacterium in human nerve cells is the Schwann cell that lies along nerve fibers in the peripheral nervous system. These Schwann cells have the optimal temperature for leprae bacteria growth at 30 degrees Celsius or 86 degrees Fahrenheit. This fairly low temperature is also why it grows on the outer portions of the body (skin) where it is cooler than deeper layers. It is a gram positive, rod-shaped bacterium that has a thick waxy coating. The coating of M. leprae aids its virulence because it is easily taken into macrophages and some dendritic nerve cells. It survives and replicates there and prevents phagosome-lysosome fusion that would otherwise destroy the bacteria.

The outer waxy coating is the reason why it takes a long time for nutrients pass through it to support such slow growth. An interesting fact about these bacteria is that it actually takes the longest period to double in population of all bacteria also known as its “incubation period.” This
time is a close to two weeks (14 days). This makes it hard to grow in a laboratory because it takes such a long time for the bacteria to multiply and initially become present in sufficient numbers. Because it has never actually been grown in a lab, M. *leprae* is instead grown on mouse foot pads and armadillos to research it further. This is the concept that illustrates why it may take years to show up in an infected individual. The waxy coating characteristic is also why carbolfuschin is used to stain it instead of the insufficient gram method. The mycolic acids in their membranes are lipids that make it hard to fight the bacterium with antimicrobial drugs because they are large and form a hydrophobic shell. The bacteria resist the dilute acid used in common decolorization procedures and appear red or violet in blue surrounding tissue (called “acid-fast” bacteria). An acid fast bacillus (AFB) smear can, therefore, be used to identify the presence of bacteria from the Mycobacterium genus but cannot confirm that M. *leprae* is specifically present. An actual AFB culture must be grown on tissue to confirm its presence or absence at that point.

**Mycobacterium Leprae Clusters (shown violet/pink)**

Under a Microscope

The most common reservoir for the bacterium that causes Hansen’s disease is in fact humans! Infected people that are not treated spread it to other people they are in close contact with (through a handshake, hug, etc.). Though armadillos have recently been found as a reservoir in the Southern United States, the effect on the statistics of infection is negligible, occurring in a minor percent of people who actually handle these armadillos. Therefore, prevention of the disease is focused on methods of earlier detection in humans to prevent its spreading. The M. *leprae* bacteria that cause HD are also known to be transmitted through droplets in the nasal mucosa or mouth when coughing or sneezing. Even though this might imply that HD is highly contagious like a cold, it is not. Close person to person contact must occur with these transmission varieties in order to contract HD. Symptoms of the disease usually do not present themselves for years because the first symptom is usually the loss of feeling, usually in the hands in feet, due to nerve damage. Ninety percent of cases that appear have numbness as the first symptom, followed by loss of temperature sensation. The final three sensations that leave are light touch, then pain, then deep pressure.

Skin lesions may not appear until years after the numbness starts. The clinical presentation of Leprosy infection has a very wide range due to the nerves and areas it affects. There is a continuum used to diagnose these that goes from the mildest “indeterminate” form to the most severe “lepromatous” type. Intermediate Leprosy (IL) is the earliest form that presents with very small numbers of hypopigmented macules (lightly colored spot or blemish) on the skin. The loss of sensation is rare. Even though most cases in this stage progress to the next
form, those with strong immune systems may clear the infection on their own or remain in this stage without going to the next one. The next phase of infection is called Turberculloid Leprosy (TT) in which the lesions are large and have loss of feeling (anesthesia) associated with them. The third phase that TT can develop into is called Borderline Leprosy (BB). In this stage lesions are still numerous but they now can take many forms including papules (solid pimple or raised swelling with visible borders), plaques (similar to papules but more flat on top and resembles a plateau), and nodules. Nodules are skin lesions in the outermost layer (epidermis), deeper layer (hypodermis), or even deeper layer (subcutaneous tissue). These nodules mean that the bacteria have gone deeper and deeper in the skin, signaling a more severe infection compared to the previous stages. And finally, Lepromatous Leprosy or “LL” is the most severe presentation of HD. Unlike the previous phases of infection, LL cannot digress back to the other stages. Loss of eyebrows and eyelashes happens as well as eye-related issues like glaucoma, pain and sensitivity to light, and blindness. Skin lesions and nodules are many. The testicles can shrink and become sterile. If the Larynx becomes infected, hoarseness of the voice will happen. And if the nose is infected, a saddle-nose deformity can occur. Health workers diagnose leprosy according to some of the main signs of the disease. These are one or more hypopigmented skin patches, one or more thickened peripheral nerve(s), and/or a positive skin smear for the bacterium. The most accurate way to determine infection is a skin biopsy in which a sample of skin is closely looked at under a microscope.

In the 1980’s, prevalence of leprosy was at a staggering 11 million cases worldwide. By the late 1990’s, this number drastically changed to a much smaller 2.5 million cases worldwide. Thankfully, by 2008 only around 249,000 cases worldwide were estimated with a very low 100 cases in the United States. The number of cases reported in the United States has stayed at about the same rate (100-200) for the past thirty years. This means the majority of cases occur elsewhere in the world. In 2002, 760,000 cases were found around the world with 90% of the cases occurring in Brazil, Madagascar, Mozambique, Tanzania, and Nepal. Seventy percent of the total numbers of Leprosy cases found worldwide occur in India.

Because Hansen’s Disease is usually carried by humans and serious cases involve human to human transfers, the best preventative measures are ones of awareness. Public education and community awareness of the signs and symptoms of the disease are important. Patients who have a confirmed or even plausible leprosy infection should be monitored closely for increasing symptoms and be handled with proper hygiene practices so as to not contract the disease in a hospital worker or family member. As mentioned earlier, there is no preventative vaccine people can take for Leprosy so people must be aware of how the disease manifests itself in people (types
of skin lesions, numbness or loss of feelings, and issues particularly affecting the hands, feet, skin, and eyes and the nerves that innervate them).

References


