Sarcoidosis

Sarcoidosis is a disease that causes inflammation of the body's tissues. It affects multiple systems and is characterized by the formation of granulomas (small lumps) that can be either inside the body or on the body's exterior. Sarcoidosis predominantly affects the lung and the lymphatic system, but virtually any organ can be involved.

In sarcoidosis, immune system cells that cause inflammation overreact and cluster together to form tiny lumps called granulomas. If too many of these granulomas form in one organ, it may not be able to work correctly. For example, if the granulomas damage healthy tissue in the lungs, scarring and stiffness may occur and limit how much air the lungs can hold. This condition is known as pulmonary fibrosis. The problems caused by sarcoidosis differ depending on which organ is affected.[1,2]

Who Has It

It is found throughout the world, among almost all races and ages and in both sexes. However, it is most common among African Americans and northern European Whites. People of Scandinavian, German, Irish, Asian and Puerto Rican origin also are more prone to sarcoidosis than the general population.[3]

Prevalence rates for sarcoidosis can only be estimated because it can easily escape diagnosis. Prevalence estimates in the United States range from less than 1 to 40 cases per 100,000 population.[4] Sarcoidosis can affect people of all ages, but it occurs most commonly in adults from 20 to 40 years of age.[5] Newer research suggests that there is a second peak in sarcoidosis, especially among women, over the age of 50.[6]

Deaths

Although uncommon, death from sarcoidosis can occur if the disease causes serious damage to a vital organ. The most common cause of mortality associated with sarcoidosis is pulmonary fibrosis resulting from the disease.[7] In the United States, there were 924 deaths due to sarcoidosis in 2006, an age-adjusted death rate of 0.32 per 100,000. Both of these numbers are higher than the average number of deaths (about 821 per year) and the age-adjusted death rate (0.30 per 100,000) for 1999 through 2006.[8]

Cause

The cause of sarcoidosis is not yet known. Most researchers agree that sarcoidosis involves an altered immune system but they do not know the source of the problem or what triggers such a

response. Some researchers believe that sarcoidosis results from a respiratory infection caused by a virus, bacteria, or an unidentified environmental toxin. There is also some evidence of a genetic basis for sarcoidosis. Current theories are that sarcoidosis develops from an interaction between a preexisting genetic risk for it and a triggering event, such as an infection or environmental exposure. More research is needed to determine the exact cause for this disease.[9]

Symptoms

In more than 90 percent of cases, sarcoidosis affects the lungs. Respiratory symptoms are present in one-third to half of cases, such as shortness of breath, dry cough, and chest pain.[10] Other common symptoms include fatigue, lymph node swelling or soreness, weight loss, and reddened, watery, or sore eyes. In some cases, symptoms can also appear outside of the lungs, such as lumps, ulcers, discolored skin or skin sores on the back, arms, legs, scalp and face.[11]

Diagnosis

Diagnosing sarcoidosis is a process of elimination. Many other respiratory diseases must be ruled out first. X-rays and other scans are often used to check the lungs and other organs for granulomas. A sample of tissue from the affected area (biopsy) is usually required to confirm the disease. When the lungs are involved, a bronchoscopy is used to acquire the tissue sample. In this procedure, a long, thin tube is inserted through the nose or mouth and down the throat to the lungs.[12]

Treatment

Treatment for sarcoidosis varies for each individual patient. In over half of the cases, sarcoidosis only lasts for 12 to 36 months.[13] In cases that do not involve certain organs or that have no additional problems from the disease, treatment is not always necessary.[14] However, 10 to 20 percent of sarcoidosis patients are left with permanent effects from the disease.[15] Among those whose lungs are impacted, 20 to 30 percent end up with permanent lung damage.[16]

For a small percentage of patients, their sarcoidosis can become chronic, lasting for many years. For those patients, therapy primarily targets ways to keep the lungs and any other affected organs working and to relieve the symptoms. Steroids are commonly prescribed to reduce inflammation. Frequent check-ups are also important so that doctors can monitor the illness and if necessary, adjust treatment.[17]

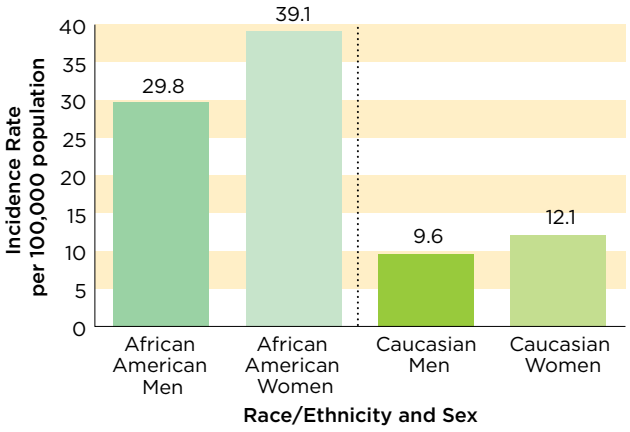
Most people with sarcoidosis can lead normal lives. Patients need to follow instructions from their physician and take all medication diligently. It is also particularly important that sarcoidosis patients do not smoke, and avoid exposure to dust and chemicals that can harm the lungs.[18]

Racial/ Ethnic Differences

African Americans

The burden of sarcoidosis is borne disproportionately by African Americans in the U.S. The age-adjusted incidence rate for African Americans is over three times that of Caucasians, at 35.5 versus 10.5 cases per 100,000 population, respectively. Incidence of sarcoidosis is consistently higher for African American females (39.1 per 100,000) compared with African American males (29.8 per 100,000), and African Americans of both genders compared to Caucasians males (9.6 per 100,000) and females (12.1 per 100,000; Figure 1). African American females 30 to 39 years of age have the highest rate of any specific age group at 107 per 100,000.[19]

Figure 1: Sarcoidosis Incidence Rate by Race/Ethnicity and Sex



Source: Rybicki BA, Major M, Popovich J Jr., Maliank MJ, Iannuzzi MC. Racial Difference in Sarcoidosis Incidence: A 5-Year Study in a Health Maintenance Organization. American Journal of Epidemiology. 1997; 145(3): 234-41.

Although sarcoidosis has a low overall mortality rate, it is more likely to be chronic and fatal in African Americans.[20] African Americans have 17 times the mortality rate due to sarcoidosis compared to Caucasians. In the United States, the mortality rate among African Americans is 1.7 per 100,000 compared with 0.1 per 100,000 among Caucasians. The age-adjusted mortality rate for African American women (1.9 per 100,000) is higher than for African American men (1.3 per 100,000).[21]

Sarcoidosis is more severe in the African American population. Those of European descent are less likely to have symptoms compared to African Americans. Sarcoidosis is also more likely to be spread throughout the body in African Americans, who show a higher frequency of ophthalmological (relating to the eyes), cutaneous (relating to the skin), hepatic (related to the liver), and lymphatic symptoms than Caucasians.[22]

A review of patients with sarcoidosis found that granulomas from African-Americans were twice as dense on average as those from Caucasian patients. When adjusted for disease stage, the granulomas from the bronchial tissue of African-American patient was 49 percent more dense than that from Caucasians, but tissue from their alveoli was only 23 percent more dense, a difference that was not significant. These differences could help explain disparities in disease severity at diagnosis between African-American and Caucasian patients with sarcoidosis.[23]

In a review of African-American sarcoidosis patients, 90 percent had a at least one other disease. Those additional diseases included high blood pressure (hypertension, 39%), diabetes mellitus (19%), anemia (19%), asthma (15%), gastroesophegal reflux disease (15%), depression (13%), and heart failure (10%). This high prevalence of additional diseases among African-Americans with sarcoidosis may

affect the prospect of survival and recovery, as well as the symptoms of sarcoidosis itself. It is also an important reason to screen for such conditions during diagnosis.[24]

African American sarcoidosis patients have higher prevalence of family history of sarcoidosis compared with Caucasian patients. African Americans with sarcoidosis are 3 times more likely than Caucasians to have a first-degree or second-degree relative with the disease.[25] In African Americans, the sibling recurrence risk ratio is approximately 2.2.[26] This means that among African Americans, brothers and sisters of someone with sarcoidosis are 2.2 times more likely to have the disease than someone in the general population. This increased risk may be due to siblings having similar environmental exposures, sharing an inherited (genetic) risk, or a combination of both these factors.[27]

Since sarcoidosis occurs more often among certain families and specific races, researchers believe there may be a genetic factor associated with its development. A scan of the entire genome among African American families, with follow-up fine mapping studies, identified chromosome 5 as a potential home for a gene that could be related to sarcoidosis risk. Follow-up studies are currently underway in order to investigate regions linked to this gene.[28]

Two common genetic markers[i] were found to be associated with increased (OR = 1.78) and decreased (OR = 0.39) risk of sarcoidosis, respectively, but only among African Americans. This genetic area is thought to affect inflammation through a chain of other actions. These findings support the idea that blacks may be at increased risk of sarcoidosis due to genetic factors.[29]

Hispanics/Latinos

Very little data are available on sarcoidosis among Hispanics and Latinos in the United States. However, looking at the incidence of this disease in the countries of origin gives an indication of the likely impact on these populations. Sarcoidosis is rarely reported in Central and South America. In Spain, the incidence rate is only 1.2 per 100,000 persons.[30]

Like Caucasians, Puerto Ricans and Mexicans are more likely to have sarcoidosis that involves the skin. Specifically, they are at risk of developing a skin condition with lesions known as Erythema nodosum. The lesions consist of raised, red, tender bumps or nodules on the front side of the legs, and nearby joints are usually sore and swollen. Erythema nodosum usually goes away within six to eight weeks. Sarcoidosis may remain undiagnosed among certain populations, including Spain, Portugal, and South America, due to a lack of screening and a greater focus on other, similar diseases that mask sarcoidosis detection, such as tuberculosis, leprosy or fungal infections.[31]

i Haplotypes -1377G/-690T/-670G and -1377G/-690C/-670A, respectively.

Asian Americans and Native Hawaiians/ Pacific Islanders

No data are available on sarcoidosis among Asian Americans/Pacific Islanders. However, looking at the incidence of this disease in the countries of origin gives an indication of the likely impact on these populations. Within these populations, sarcoidosis primarily affects Japanese people. The disease is rare in Southeast Asian, Korean, Chinese, and Indonesian populations.

In Japan, the annual incidence of sarcoidosis ranges from 1 to 2 cases per 100,000 people.[32] Sarcoidosis occurs most often in both Japanese men and women between the ages of 25 and 40 years.[33]

In Japanese sarcoidosis patients, cardiac and ophthalmological symptoms are common. Cardiac involvement is most common in females over the age 50, compared with Europeans and Americans.[34] Death due to heart complications is also much more common in Japan than the U.S., accounting for 77 percent of deaths related to sarcoidosis there.[35]

Japanese sarcoidosis patients sometimes face a different set of obstacles than Caucasian patients. In a Japanese study of 228 sarcoidosis patients, 8.8 percent were found to have airflow limitation, and none had airway reversibility. Unfortunately, airflow limitation in patients with sarcoidosis is associated with poor prognosis.[36]

American Indians/ Alaska Natives

There are no data available on Americans Indians/Alaska Natives.

Resources

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