Sarcoidosis: A Multisystem Granulomatous Disease

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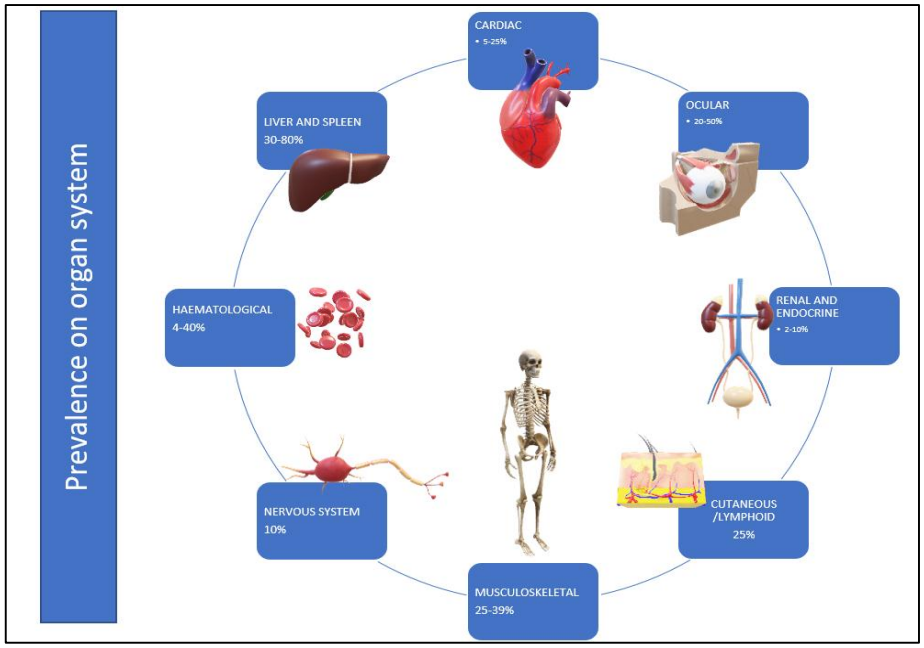
Abstract: Multisystem disease, Sarcoidosis causes the formation of lumps (granulomas) in various organs, including the lungs, liver, skin, lymph nodes, eyes, and spleen. The pathology of sarcoidosis is complicated by a number of factors, including genetic predisposition, environmental factors, and infection. Early diagnosis and identification are complicated due to the uncertain aetiology. The epidemiology, causes, organ effects, screening, and diagnosis are all discussed in this study.

Keywords: sarcoidosis, cause, inflammation, prevalence, epidemiology

INTRODUCTION

Sarcoidosis is a disease marked by inflammation in multiple systems, where granulomas or groups of inflammatory cells form in different organs, causing inflammation. The condition can manifest as a distinct phenotype of "sarcoidosis," influenced by the specific triggering exposure and underlying genotype (Westney, 2006). The inaugural case was documented by Jonathan Hutchinson, a physician at King's College Hospital in London, in 1877 (Hutchinson, 1877). In 1956, the Ist case of sarcoidosis in India was identified in the Journal of the School of Tropical Medicine, Calcutta, and two sisters with familial sarcoidosis were registered in 1923. (Gupta, S.K. 2002). Sarcoidosis is characterised by bilateral hilar lymphadenopathy and pulmonary reticular opacities. Skin, eyes, and joints are other major involved areas, though it can also be found in the musculoskeletal system, heart, exocrine glands, kidney, reticuloendothelial system, and CNS to varying degrees (Ma Y, Gal et al, 2018, Landi C, Carleo A,et al, 2018, Petek, Rosenthal et al, 2018).



Epidemiology

These studies are being conducted to identify the prevalence of this disease in a population and to look at risk factors. Owing to the difficulties in determining the number of asymptomatic patients, globally prevalence and incidences of sarcoidosis are unknown. Sarcoidosis affects people of all the ages, races, and ethnicities. Patients with sarcoidosis are typically between the ages of 20 and 39, and it is more common in non-smokers, women, and people who live in rural areas. (Peros, T.; Ljubic,1995).

From 2009 to 2013, a study of sarcoidosis in the United States found that the and prevalence and incidence of this disease is substantially higher in

black people (17.8 and 141.4 per 100,000) than in white people (8.1 and 49.8), with black females (178.5) having the highest prevalence (Baughman, Field et. Al 2016). Sarcoidosis is notably widespread in India, as evidenced by a respiratory unit in western India recording 10–12 cases per 1000 new registrations, and a respiratory unit in the capital region reporting 61.2 cases per 100,000 (Sharma, S.; Mohan, 2004). Hyperlipidaemia, obesity, diabetes, thyroid disease, coronary heart disease, osteoporosis, asthma, chronic renal disease, hypertension, and COPD are the most common co-morbidities seen in sarcoidosis patients (Martusewicz, M., et. Al 2015, Nowinski, A.,et.al 2014). Patients with autoimmune disorders such as autoimmune thyroid disease, ankylosing spondylitis, and systemic sclerosis are often diagnosed with sarcoidosis (Carmi, O., et. Al 2018). In conclusion, sarcoidosis is more prevalent among women than men, and individuals of African-American descent are at a higher risk for these conditions. People with a sarcoidosis family background are at a far greater risk of developing the disease. Adults of age between 20 and 40 are most affected, and children are rarely affected.

Cause

The exact origin of sarcoidosis remains unidentified. Numerous investigations have suggested that a blend of hereditary predisposition, environmental elements, and autoimmunity may contribute to the development of this disease, yet a definitive single cause of this disease has not been identified yet.

Genetical factors. As discussed in epidemiology genetic factors also have crucial role in causing sarcoidosis. To date, 11 sarcoidosis risk loci have been identified: HLA-B, HLA-DPB1, IL23R, SH2B3/ ATXN2, ANXA11, RAB23, bIL12B, NFKB1/MANBA, BTNL2, FAM177B, chromosome 11q13.1, and (Fischer, A.,et. Al, 2015).

According to genome-wide association studies, the development of this disease is linked to several non-HLA and HLA alleles (Schurmann, M.;et al,2001). Sarcoidosis susceptibility is associated with significant markers such as HLA-DRB10301/ DQB10201, transforming growth factor β (TGF- β) (Pabst, S. et al., 2011), TNF -α (Sharma, S. et al., 2008), and TLR-4 (Pabst, S. et al., 2006), as highlighted in studies by Grunewald, J. (2010) and Iannuzzi, M.C. (2007).

Environmental risk factors: An elevated risk of this disease in the environment is associated with factors like soil, wood stoves, inorganic particulates, tree pollen, nanoparticles, and insecticides. and. Furthermore, specific occupations, including those in gardening, hardware, metalwork industries, and building supplies, along with navy ship servicemen, educators, and firefighters, are also prone to sarcoidosis (Newman, K.L.; et. Al 2012). An elevated risk of sarcoidosis has been associated with silica toxicity, as indicated by Vihlborg et al. in 2017. The underlying hypothesis for this connection is that environmental factors play a substantial role in the progress of sarcoidosis. This notion is supported by evidence showing an increased risk of sarcoidosis among US World Trade Center staff exposed to the crash debris, particularly firefighters, as demonstrated by Izbicki et al. in 2007.

Infections. Microbial agents such as Leptospira species, Mycoplasma species, herpes virus, retrovirus, Chlamydia pneumoniae, Pneumocystis jirovecii, Borrelia burgdorferi (Newman, L, 2005), Propionibacterium species and Mycobacterium(M.tb) (Drake, W.P. et al. 2006), have been identified in abundance in studies (Ishige, I. et al,2005). Patients who received interferon therapy for hepatitis C infection developed ed sarcoidosis, according to testimony (Hirano, A et al,2005, Trien, R.et al,2014). According to some reports, C type of hepatitis infection can increase the risk of developing sarcoidosis on its own. Interferon treatment, on the other hand, appears to increase interferon- and interleukin-2 expression, causing granuloma formation and hence sarcoidosis (Brjalin, V,et al, 2005, Ramos-Casals et al, 2005). Autoimmunity is a condition in which a person's immune Sarcoidosis may occur in conjunction with other rheumatoid arthritis, autoimmune diseases, Sjogren's syndrome, ankylosing spondylitis, autoimmune thyroid disease, and are among the conditions (Korsten, P., et al,2018). Previous research has extensively examined the role of the microbiota in autoimmune diseases, demonstrating its significant contribution to their pathogenesis (Chu, F., et al, 2018). Consequently, exploring the microbiome of individuals with sarcoidosis and its connections to other diseases could present novel opportunities for investigating the underlying causes of the condition (Becker, A., et al, 2019; Inaoka, P.T., et al, 2019).

Sarcoidosis on organs

On lungs

Most commonly affected organs on body with 90% prevalence. Symptoms of sarcoidosis in lungs includes:

- Coughing that is dry
- Problem in breathing, wheezing, and pain with breathing
- Pain in chest or pressure, tightness, or distress
- Blood in cough (an unusual symptom), especially in the early stages of sarcoidosis.

Untreated sarcoidosis can lead to many long-terms diseases like fibrocystic disease, pulmonary fibrosis, pulmonary hypertension, aspergilloma, bronchiectasis.

Lymph nodes

The lymph glands in the chest are commonly the most affected in this condition, but they're difficult to spot on an x-ray. Neck, under the chest, armpit, and groyne are several other areas where lymph can swell.

Skin

Sarcoidosis causes problems in skin like

- rashes usually on ankles,
- change in color of skin (somewhere darker somewhere lighter) ,
- nose, cheeks, and ear lesions,
- nodule formation.

Eyes

Sarcoidosis affects eyes without showing symptoms but if symptoms occurs they cause

- extreme redness,
- distorted vision,
- pain in eyes,
- burning, scratching, or dry eyes;
- light sensitivity.

Problems that are associated with sarcoidosis in eyes are: ubieties, dry eye syndrome, tear gland enlargement, glaucoma, cataracts, blindness.

Heart

In US more than 10 % people are affected by the sarcoidosis of heart. Patients which have symptoms includes:

- Pain in chest
- Breathing problems (dyspnoea)
- You're fainting (syncope)
- Tiredness
- Heartbeats that are irregular (arrhythmias)
- Fluttering or rapid heartbeat (palpitations)
- Oedema (swelling caused by an excess of fluid)

Disease associated with heart sarcoidosis are Heart blocks, arrhythmias, Heart failure, Heart valve problems, Pericarditis, Heart attacks.

The nervous system and the brain

- Sarcoidosis is a disease that affects the nervous system and brain, with a prevalence of 15%. Symptoms of the brain and nervous system include:
- Facial palsy (also known as Bell's palsy) is a condition in which one side of the face droops and/or becomes paralysed (the most common nervous system symptom)
- Migraines
- Redness and pain in the eyes
- Weakness, numbness, tingling, and/or pain in the ears, arms, and/or legs
- Blindness
- Seizures
- leg or Arm paralysis
- Mood and behaviour changes, memory loss, irritability, and hallucinations (which are rare)
- Disease associated with sarcoidosis of brain and nervous system:

Inflammation and/or damage to the nerves Peripheral neuropathy, also known as peripheral neuropathy, is a condition that affects Meningitis, hydrocephalus, neuroendocrine disorders, and coma are all symptoms of granulomas in the meninges.

Kidney and urinary tract

Prevalence of these in human are 2 to 10 % Symptoms which are included are due to the development of kidney stone like increased urge to pee, pain in back or sides. Problems associated with the sarcoidosis are increased level of calcium in blood and urine, kidney stone and kidney failure.

Screening and diagnosis

Sarcoidosis is diagnosed using radiological and standard clinical evidence after histological confirmation of narcotic granulomas. And patient undergoes multiple examination, according to the involvement of organ. All examinations according to symptoms mentioned above are used to identify

sarcoidosis because there is no standard establishment yet. To confirm the presence of sarcoidosis one or 2 more tests can be performed, but history, symptoms and other signs are enough for the relevant medical examination.

CONCLUSION

Despite the twenty-first century and ongoing studies on sarcoidosis, the aetiology of sarcoidosis remains an open question. When these factors interact with other environmental factors, the prevailing theory underscores the substantial impact of host-microbe interaction and genetic factors on the development of this illness. Clinicians are in immediate need of effective diagnostic techniques to identify and predict the progression of sarcoidosis for the purpose of finding a cure.

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