COMPLEMENT DEFICIENCIES

When the Immune System has Inadequate Levels of Complement Proteins

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Complement deficiencies can contribute to recurrent infection and they can contribute to autoimmune disease development, particularly immune complex disorders.

Deficiencies of Complement

Complement deficiencies include several disorders caused by decreased levels of specific complement proteins. Complement is necessary for the immune system’s functioning, particularly the destruction of infectious agents. Without adequate complement, the body has increased susceptibility to infection and other illnesses, including certain autoimmune diseases.

Who is Affected?

Both sexes are affected equally. Individuals with C3 deficiency usually develop symptoms in childhood, while MAC deficiencies affect young adults and adolescents. Deficiencies that contribute to lupus such as C1, C2, and C4 usually develop in young adulthood through middle age. However there is no clear pattern of age at first presentation.

Causes

Complement deficiencies can be genetic occurring as a primary deficiency. Or they can be secondary, caused by ailments that involve protein loss, including severe burns, liver or kidney diseases, malabsorption disorders such as celiac disease, and systemic autoimmune diseases such as systemic lupus erythematosus (SLE). In autoimmune diseases, adequate complement is needed to prevent the formation of immune complexes. Immune complexes are particularly destructive in their ability to lodge into kidney tissue, causing glomerulonephritis.

Symptoms

Symptoms of complement deficiency depend on the specific complement deficiency and the disease caused by the deficiency. Without adequate complement, the body is prone to frequent infections, such as pneumonia and meningitis. Because there are approximately 30 different forms of complement, the specific symptoms depend on the complement component that’s lacking.

Some people with complement deficiencies remain healthy while others develop frequent fevers, headaches or cough. In people with autoimmune disorders such as systemic lupus,
complement deficiency can cause weight loss, joint pain, and rash. Other autoimmune conditions related to complement deficiency include hereditary angioedema, paroxysmal nocturnal hemoglobinuria, or leukocyte adhesion deficiency syndrome. These disorders can cause edema or swelling of the face, skin infections or abdominal and back pain.

Diagnosis

Blood tests are used to measure total complement levels using the CH50 and the APH50 tests. Specific tests can also be used to measure levels of C1, C2, C3, C4 C1q deficiency and others. Individuals with C1q deficiency are reported to have a 93 percent chance of developing systemic lupus erythematosus. C1rs is associated with a 57 percent chance of developing systemic lupus, and C4 deficiency is associated with a 75 percent chance of developing lupus. New tests for membrane attack complex (MAC) components such as C5b-9 are also available. Deficiencies of C3 result in recurrent pyogenic infections.

Individuals with MAC deficiency are at particular risk for infection with Neisseria gonorrhoeae, which causes gonorrhea and Neisseria meningitidis, the causative organism in bacterial meningitis.

Treatment

There are no blood transfusion products available to replace complement. Treatment is usually preventive with antibiotics used at the first sign of infection, and vaccines used to prevent specific disorders such as flu.

Resources:

R. Krishna Chaganti, Complement Deficiencies, eMedicine from WebMD, June 2, 2006,


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