

IMMUNOLOGY

COMPLEMENT DEFICIENCY IN SYSTEMIC AUTOIMMUNE DISEASES

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Abstract: Complement is an important part of the innate immune pathway. It involves over 20 serum proteins, most being synthesized in liver. These proteins are initially inactive precursors which get activated later by different stimuli. All the three pathways of complement activation i.e., classical, alternative and lectin converge to produce membrane attack complex or terminal complex which leads to lysis of the target pathogen. Activity of complement is controlled by regulatory proteins that prevent host cell damage and lysis caused by inadvertent binding of activated complements. Complement deficiency results in autoimmune diseases. Early complement deficiency results in monogenic lupus and infections due to encapsulated bacteria whereas late complement component deficiency causes neisserial infections. Complement can be assessed by various tools like enzyme-linked immunoassays, flow cytometry, and next-generation sequencing.

Keywords: Early-onset systemic lupus erythematosus, Complement, Classical pathway, Autoimmune diseases, Atypical hemolytic uremic syndrome, Alternative pathway.

Points to Remember

- *Complement plays a key role in pathogenesis of autoimmune and inflammatory diseases.*
- *Monogenic lupus can be due to defects in components of classical pathway.*
- *Complement deficiency results in a predisposition to infections primarily from encapsulated bacteria.*
- *Defective regulation of complement system can result in atypical hemolytic uremic syndrome or paroxysmal nocturnal hemoglobinuria.*
- *Excessive alternative pathway activation can cause lupus nephritis or antiphospholipid antibody syndrome.*

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