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- Phagocytic AIHA is termed extravascular, whereas complement-mediated lysis of RBCs is termed intravascular AIHA.
 - In order for intravascular AIHA to be recognizable, it requires overwhelming complement activation, therefore most AIHA is extravascular – be it IgG- or IgM-mediated.
 - Each IgM molecule has the potential to activate a C1 molecule so that large amounts of complement are found on the RBCs in cold agglutinin disease. These sensitized RBCs may undergo intravascular complement lysis or may be destroyed in the liver.

CLINICAL FEATURES

➤ HX:

- Recent infection, immunological dx

➤ SIGNS/SYMPTOMS:

- Non-specific fever,
- weakness,
- fatigue,
- Shortness of breath,
- jaundice,
- pallor,
- dark urine
- Others related to the aetiology as in secondary AIHA

TREATMENT

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SYMPTOMATIC

- Blood Transfusion

1ST LINE:

- Prednisone
- Methylprednisolone

2ND LINE:

- Rituximab
- Splenectomy
- Danazol

3RD LINE:

- Azathioprine
- Cyclophosphamide
- Mycophenolate

- The peak incidence was in the first 4 years of life. Predominance among males was noted. Two familial cases were observed. Two main clinical patterns were distinguished: an acute transient type and a prolonged chronic type. A statistically significant correlation was found between the presenting clinical, hematologic and immunologic features and the ultimate evolution of the disease into either clinical pattern. High titer cold agglutinins were very rarely observed. In no patient was an underlying malignant disease found.

REFERENCES

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