REVIEW ARTICLE

Asherson's Disease

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Abstract

Asherson's disease is a rare autoimmune disorder. It affects the anticoagulation mechanism of blood. The exact cause of the disease is unknown but it may be due to antiphospholipid (APL) syndrome, hereditary, and gene transformation. This disease may affect all organs in the human body. There is no cure for the disease and no exact medicine has been identified. Supportive treatment is given to promote the betterment of the individual with Asherson's disease.

Keywords: Antiphospholipid syndrome, Asherson's disease, Coagulation factor, Hereditary.

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Introduction

Asherson's syndrome is an extremely rare autoimmune condition which is shortly known as catastrophic antiphospholipid syndrome (CAPS). It is defined as the there is a fast progression of blood clots affecting many organs within an hour, days, or weeks. The major triggering factor of the disease may be due to the administration of vaccination, or wounds due to any physical injury. Infection is caused by microorganisms and the body's anticoagulation mechanism failure. It is also defined as the presence of certain antibodies [antiphospholipid (APL) antibody] which is helpful for the formation of blood clots in the body.[1]

An individual who had defects in their anticoagulation mechanism which is due to recurrent injury or bleeding are more prone to get this syndrome. Individuals with previous experience of an episode of the APL syndrome will easily get Asherson syndrome.[1]

Incidence

Ashersons's disease has been identified since 1992, around 300 people were affected by this condition, and women are more prone to get affected when compared to men. This can affect at any age-group, but the majority of the affected people are in the younger age-group.

Etiological Factors

- All autoimmune disorders
- History of genetic and environmental factors
- The cause of the syndrome is uncertain
- Infection is the primary cause
- History of primary or secondary APL syndrome
- Injuries due to invasive surgical operations, the discontinuation of anticlotting medication, pregnancy, and some underlying malignancies
- Additional factors like cancers[2]

Classification

- Systemic inflammatory response syndrome (SIRS): Development of within a short period (less than a week), multiorgan failure, small vessel thrombosis, abnormal thrombosis of bone marrow, and reproductive organs[3]
- Development of laboratory signs of APL syndrome (APS)[4]

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Pathogenesis

Due to etiological factors (genetic thrombophilia, poor APL-in-circulation) and precipitating factors (malignancy, infections, obstetric complications, surgery, trauma, withdrawal of anticoagulant medications, oral contraceptives)

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Activation of coagulation cascade (abnormal cellular proliferation and differentiation impacting on cell function)

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Inflammation is a central pathogenic factor in APS

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Disseminated intravascular coagulation, SIRS

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Systemic microvascular thrombosis

↓

CAPS (Asherson's syndrome)

↓

Multiple organ failure[5,6]

Clinical Features

- Development of numerous blood clots (thromboses) within a short period of time

- Depending on the organ system involvement symptoms may differ[7]
  - Renal: Oliguria and hypertension
  - Cutaneous: Livedo reticularis—blotchy reddish discoloration of the skin, gangrene, and bruising
  - Neurological: Stroke (cerebral infarction), seizures, encephalopathy[8]
  - Cardiovascular: Angina, myocardial infarction, valvar heart disease, mitral valve regurgitation (MVR)
  - Gastrointestinal: Abdominal cramping and pain[9]
  - Hematological: Bone marrow (anemia, thrombocytopenia)
  - Endocrine: Hormone imbalance and low blood pressure[10,11]

Diagnosis

- Enzyme-linked immunosorbent assay (ELISA) is used to detect anticardiolipin antibodies in the blood
- Multiple clots have been identified in at least three different organs within a week
- A coagulation test is used to detect the presence of lupus anticoagulation in the blood[8]

Complications

- Renal: Decreased blood flow causes renal failure
- Neurological: Decreased blood flow to the brain causes stroke, partial paralysis, and loss of speech
- Cardiovascular: Necrosis and gangrene formation due to blood clots in the valve of the vein and cardiomegaly, and cardiac failure.
- Pulmonary: Pulmonary hypertension and pulmonary embolism
- Obstetrics: Hemolysis, elevated liver enzymes, low platelet count (HELLP) syndrome, preeclampsia, miscarriages, stillbirth, intrauterine growth retardation (IUGR), endothelial dysfunction, ischemia, and placenta microthrombosis and placental abruption
- Rarely, in some severe cases, APL syndrome can cause multiple organ damage in a short period of time medical management[9]

Medical Management

There is no conventional treatment. Some treatment regimens are:

- Anticoagulants: Heparin is administered intravenously corticosteroids are used to reduce necrosis
- Sometimes steroids and heparin are given together to have a better effect
- Antibiotics: Infections are treated by intensive antibiotic therapy
- Immunoglobulin (specific proteins), and plasma exchanges are done by plasmapheresis method
- Monoclonal antibodies: Administration of rituxan and eculizumab in high doses to the affected individual

Dietary Management

Foods to Include

- Green leafy vegetables: These are rich in magnesium which is an essential nutrient for autoimmune disease (e.g., Spinach, broccoli)
- Turmeric: It has anti-inflammatory properties and is rich in antioxidants
- Fish: Fish like salmon are rich in omega-3-fatty acids and antioxidants which help in fighting diseases.
- Berries: Berries are rich in high antioxidants. Cauliflower: It contains sulphur which fights against cellular damage and provides protection

Don't Include

- Some foods act as an aggravating factor for this disease. So it is essential to know about the foods that interact to aggravate the symptoms
- Some of the foods are:
  - Foods rich in vitamin K such as avocadoes, beans to be avoided
  - Cranberry juice has an effect on blood thinning. So it should be avoided
  - Avoid consumption of alcohol

Nursing Management

- Obtain a complete history of thrombotic events
- Assess the treatment regimen for APS based on the patient clinical condition
- Routine prothrombin time in international normalized ratio (PT/INR) to be monitored
- Eliminate the risk factors like oral contraception, smoking, alcohol, and high lipid intake
- Recurrent deep vein thrombosis (DVT) care to be done
- Instruct the patient o avoid vitamin K rich foods
- Avoid heavy exercises and sports activities if patient is on warfarin therapy
- Limit activity in patient with DVT
- Educate the patient to avoid prolonged immobilization

Conclusion

Ashesrson's syndrome is a life-threatening condition which is characterized by diffuse vascular thrombosis, results in multiple organ failure within a short period of time.[5,6] Even though the number of cases reported of this condition is low, but the mortality rate is around 50%. Because of the high rate of mortality, this disease is considered to be a serious illness. So there should be greater awareness and early diagnosis and treatment to be carried out. APL mediated.[5]

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