ORIGINAL RESEARCH PAPER
Volume - 9 | Issue - 8 | August - 2020 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

EVANS SYNDROME

Nursing

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KEYWORDS

DEFINITION:

Evans syndrome may be a very rare autoimmune disease during which the system destroys the body's red blood cells, white blood cells and/or platelets. Affected people often experience thrombocytopenia (too few platelets) and Coombs' positive hemolytic anemia (premature destruction of red blood cells). Signs and symptoms may include purpura, paleness, fatigue, and light-headedness. The exact cause of this condition is unknown. The best treatment options for Evans syndrome depend on many factors, including the severity of the condition; the signs and symptoms present; and each person's response to certain therapies. (1)

Causes:

The exact, underlying cause of Evans syndrome is unknown. Evans syndrome is an autoimmune disorder. It occurs when the immune system produces antibodies that mistakenly attack healthy tissue, specifically red blood cells, platelets and sometimes certain white blood cells.

The immune system normally responds to foreign substances by producing specialized proteins called antibodies. Antibodies work by destroying foreign substances directly or coating them with a substance that marks them for destruction by white blood cells. When antibodies target healthy tissue they may be referred to as autoantibodies. Researchers believe that a triggering event (such as an infection or an underlying disorder) may induce the immune system to produce autoantibodies in Evans syndrome.

Evans syndrome may occur in combination with another disorder as a secondary condition. Secondary Evans syndrome can be associated with other disorders including autoimmune lymphoproliferative syndrome (ALPS), lupus, antiphospholipid syndrome, Sjogren's syndrome, common variable immunodeficiency, IgA deficiency, certain lymphomas, and chronic lymphocytic leukemia.(2)

Pathophysiology

The exact pathophysiology of Evans syndrome is unknown, although the mechanism generally seems to be related to the one associated with autoimmune hemolytic anemia and immune thrombocytopenia. In these disorders, non–cross-reacting autoantibodies are directed against antigens specific to RBCs, platelets, or neutrophils.

Wang et al demonstrated decreased serum levels of immunoglobulin G (IgG), immunoglobulin M (IgM), and immunoglobulin A (IgA) in these patients. The cytopenias that occur with Evans syndrome may be related to T-cell abnormalities; decreases in helper T cells and increases in suppressor T cells were noted in these patients.

Savasan et al observed that more than half of the patients with Evans syndrome had evidence of lymphoid hyperactivity.

Teachey et al demonstrated that more than half (58%) of patients with Evans syndrome might have autoimmune lymphoproliferative syndrome (ALPS), a novel finding with potentially important therapeutic implications. Several cases of Evans syndrome have also been reported in association with the group of lymphoproliferative disorders known as Castleman disease.

Programmed cell death (apoptosis) of activated lymphocytes is critical to immune homeostasis. The cell surface protein Fas (CD95) and its ligand play a pivotal role in regulating lymphocyte apoptosis, and defective expression of either Fas or Fas ligand results in marked overaccumulation of mature lymphocytes and autoimmune disease in mice. Some study results suggest that defective lymphocyte apoptosis caused by mutations of the *FAS* gene can result in severe ALPS in humans.

Teachey et al screened 12 children by using flow cytometry for CD4/CD8 (double-negative) T cells and using the definitive test for ALPS (ie, defective in vitro Fas-mediated apoptosis). Six patients had elevated numbers of double-negative T cells and defective Fas-mediated apoptosis, and 1 had a borderline elevation; thus, 7 patients with Evans syndrome had evidence suggestive of ALPS, which, in turn, suggests that there may be some overlap between Evans syndrome and ALPS. This may explain the severe clinical course in some patients with Evans syndrome.

A study by Rivalta et al indicated that in children with Evans syndrome, the disorder is frequently the initial expression of an immunologic or rheumatologic disease. Out of 12 pediatric patients with Evans syndrome, five (42%) were found to have a primary immunodeficiency, including three with common variable immunodeficiency, one with ALPS, and one with Rubinstein-Taybi syndrome. Of the three patients (25%) with a rheumatologic disorder, two had systemic lupus erythematosus, and one had mixed connective tissue disease. (3)

Signs & Symptoms

Due to the system attack that Evans syndrome causes, people with this condition have abnormally low levels of blood cells. Doctors may refer to this as cytopenia.

Cytopenia causes a variety of symptoms that vary from person to person. The severity of the disease and therefore the specific blood cells that the disease involves will determine what symptoms an individual develops.

Some people have periods of remission during which symptoms of the disease get away. Remission may happen after successful treatment. However, people may also have periods when symptoms get worse.

Anemia Symptoms

Low levels of red blood cells thanks to Evans disease may cause anemia. Anemia means that the body does not have enough red blood cells to get oxygen to the organs and tissues.

Anemia may cause the following symptoms:

fatigue
pale skin
dizziness or lightheadedness
shortness of breath
rapid heartbeat
yellowing of the skin and eyes (jaundice)

Low Platelets

Low levels of platelets may cause bleeding under the skin because the blood cannot clot properly.

If this occurs, a person may notice:

- tiny purple or red spots on the skin (petechiae)
- purple spots on the skin, which can appear raised (purpura)
- larger purple blotches on the skin (ecchymosis)
- skin that bruises easily

Low Neutrophils
People who have low levels of neutrophils are more likely to get infections. Their symptoms may include:
- a fever
- feeling unwell
- sores or ulcers inside the mouth
- frequent illnesses

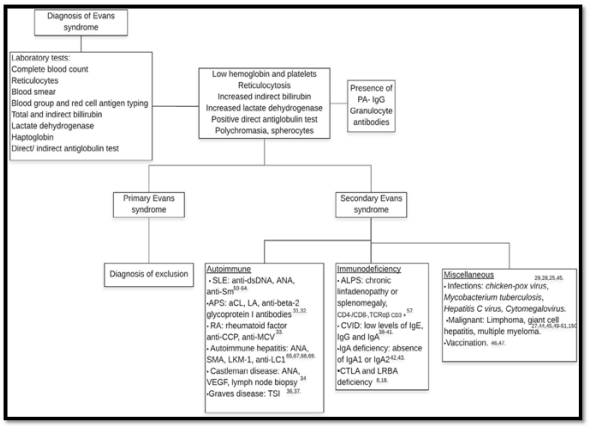
People with Evans syndrome can also have an enlarged spleen, liver, or lymph nodes.

A person may not notice these signs, but a doctor can identify them during an exam or on an imaging test. (4)

Affected Populations
The incidence and prevalence of Evans syndrome is unknown. The disorder can affect children or adults.

Diagnosis
Laboratory studies which will be considered include the following:
- Complete blood count (CBC)
- Reticulocyte count
- Coombs test (direct antiglobulin test)
- Tests for antineutrophil, and antiplatelet antibodies
- Lupus antibody (lupuslike inhibitor) and antinuclear antibody (ANA) tests
- Measurement of serum immunoglobulins
- Flow cytometry of blood samples
- Gene mutation studies

Bone marrow aspiration helps reveal aplastic anaemia or an infiltrative disorder. It is usually indicated for excluding infiltrative processes in patients who present with pancytopenia.(5)



Abbreviations:
aCL, anticardiolipin; ALPS, autoimmune lymphoproliferative syndrome; ANA, antinuclear antibodies; anti-CCP, anti-cyclic citrullinated peptide; anti-LC1, anti-liver cytosol antibody; anti-MCV, anti-mutated citrullinated vimetin; anti-Sm, anti-Smith antibody; APS, antiphospholipid syndrome; CVID, common variable immunodeficiency; LA, lupus anticoagulant; LKM-1, liver kidney microsomal type 1 antibodies; PA-IgG, platelet-associated IgG; RA, rheumatoid arthritis; SLE, systemic lupus erythematosus; SMA, smooth muscle antibody; TSI, thyroid stimulating immunoglobulin; VEGF, vascular endothelial growth factor.

Treatment And Management
There is no cure for Evans syndrome, so treatment focuses on relieving symptoms and increasing blood corpuscle counts.

Treatment for Evans syndrome varies from person to person. The most suitable treatment will depend upon the person's:
- age
- health status
- symptoms
- blood cell count

Treatment Options Can Include:
Corticosteroids

One of the foremost common treatments for Evans syndrome is corticosteroids. These medications can help lower the activity of the immune system.

However, Corticosteroids Can Have Many Side Effects, Including:
- high blood pressure
- weight gain
- mood swings
- high blood sugar

Intravenous immunoglobulin

Intravenous immunoglobulin (IVIg) is a solution that contains antibodies from healthy donors.

A person receives this treatment intravenously, which suggests through an injection into the veins. IVIg can help prevent the immune system from attacking the body.

Immunosuppressive drugs
These medications work by lowering the level of activity of the immune system. Possible side effects include a better risk of getting illnesses and infections.

Rituximab
Rituximab is a newer medication that avoids some of the side effects of immunosuppressive drugs.

Spleen removal
In some cases, treatment may involve surgically removing the spleen in a procedure called a splenectomy.

Hematopoietic stem cell transplantation
A somatic cell transplant could help reset the system and supply a possible cure for Evans syndrome. The treatment requires a donor to provide stem cells. The donor will usually be a close family member who is a proper match. However, stem cell treatment may not work for everyone, and no large studies have yet proved that it is effective. (4)

Additional potential supporting interventions
Anecdotic reports suggest that immunological benefits are often gained in patients with autoimmune diseases including SLE and atrophic arthritis from simple measures, including oral honey drink and black caraway oil152 or powder as nutritional support measures. It is reported that wet cupping therapy (Al- Hijamah) can clear patient's blood from autoantibodies, inflammatory mediators and immunological noxious substances.

Perspectives in ES
New treatment approaches for patients affected by ES should led to long-lasting remissions, less-severe relapses and an increased opportunity for cure. These scenarios are going to be possible as knowledge of the molecular landscape during this intriguing disease deepens, thus allowing the planning of sophisticated biological therapy and/or improved HSCT strategies for severe cases, particularly in children. A Phase II active clinical trial (Identifier: NTC00392951)

recently evaluated sirolimus at 3 mg/m2 then at 2.5 mg/ m2 counting on its serum levels in refractory autoimmune cytopenias including ES. A total of 30 patients were evaluated, including 8 with ES. There were three complete responses, three partial responses and two patients with no response. Main objectives were to document toxicities of sirolimus, including mucositis, elevation of triglycerides and cholesterol and to assess the efficacy of this approach in patients with refractory ES. (6)

Differential Diagnosis
- Autoimmune lymphoproliferative syndrome (ALPS)
- Thrombotic thrombocytopenic purpura (TTP)
- Systemic lupus erythematosus (SLE)
- Antiphospholipid syndrome (APLA)
- Sjogren syndrome
- Common variable immunodeficiency (CVID)
- IgA deficiency
- Lymphomas
- Chronic lymphocytic leukemia (CLL) (7)

Complications
Potential Complications Of Evans Syndrome Include The Following:
- Hemorrhage with severe thrombocytopenia – The national survey reported hemorrhage in 29% of patients, with 2 deaths resulting

from severe GI bleeding and a third death from acute intracranial bleeding

- Serious infection in patients with neutropenia – The national survey showed invasive infections in 29% of patients, including pneumonia, sepsis, meningitis with *Streptococcus pneumoniae,* localized abscess, and osteomyelitis ; one patient died of presumed sepsis and liver failure 9 years after splenectomy

Genetic Counseling

Most of the cases are sporadic. Familial cases have exceptionally been observed, mainly in the setting of an underlying primary immunodeficiency. (8)

Prognosis

In a nationwide study of Evans syndrome the median survival was 7.2 years (primary Evans syndrome: 10.9 years; secondary Evans syndrome: 1.7 years). Secondary Evans syndrome was associated with higher mortality rate than primary Evans syndrome, with a 5-year survival of 38%. Among patients with Evans syndrome, the prevailing causes of death were bleeding, infections, and hematological cancer. (9)

Summary

Evans syndrome is a rare immune system disorder that reduces a person's blood cell count and can lead to anemia and other complications.

Although there is no cure, newer treatments and advanced medical care can help relieve symptoms and improve quality of life.

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