Evans syndrome

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Description [edit]

Evans' Syndrome is a combination of two conditions: autoimmune hemolytic anemia and autoimmune thrombocytopenia purpura. Autoimmune hemolytic anemia is a condition in which there are low levels of iron in the body due to the destruction of the red blood cells that normally carry oxygen. Autoimmune thrombocytopenia is revealed by a low level of platelets in the blood due to their destruction in the circulation. Platelets are a component of blood that is responsible for creating clots in the body to heal wounds.

Those Affected [edit]

The incidence of Evan's Syndrome is not precisely known. The syndrome is reported to be a complication affecting 4-10% of those persons with a particular type of thrombocytopenia known as autoimmune thrombocytopenia purpura. The syndrome is more prevalent in children than in adults.

Signs and Symptoms [edit]

The signs and symptoms of Evan's Syndrome will be a combination of the signs and symptoms of the two underlying conditions. In autoimmune thrombocytopenia purpura the following may be found: Bleeding of skin or mucus lined areas of the body. This may show up as bleeding in the mouth, or purpuric rashes (look almost like bruises), or tiny red dots on the skin called petechiae. Laboratory results will show low levels of platelets

In autoimmune hemolytic anemia the following may be found: Fatigue, Pale skin color, Shortness of breath, Rapid heartbeat, and/or Dark urine

Possible Causes [edit]

The cause of the signs and symptoms of Evan's Syndrome are directly related to the low levels of red blood cells (RBC) and platelets in the blood. These low levels are a result of circulating antibodies that bind to the blood cells and destroy them. Antibodies are made under normal conditions against foreign substances in the body and are therefore very useful in warding off infection. In conditions that are referred to as "autoimmune" the body makes antibodies against itself. In the case of Evan's Syndrome, it is not currently known what triggers this reaction to happen.

Diagnosis [edit]

The diagnosis of Evan's Syndrome is based primarily on laboratory findings, as well as the corresponding physical signs and symptoms. A complete blood count (CBC) will confirm the presence of anemia and low platelets. Additional studies may include a peripheral smear and a Coombs test. A peripheral smear of the blood may reveal evidence of red blood cell destruction or reticulocytosis. Reticulocytes are immature red blood cells and are usually abundant in Evan's syndrome where there is a need to replace ongoing losses. There are also distinct shapes to certain cells that may be found when a sample of the patient's blood is viewed under a microscope. In patients with Evan's syndrome the red blood cells may appear small and globular shaped (then called spherocytes) but will not be fragmented. A Coombs test is used to detect the presence of antibodies against the RBC and is usually positive in immune mediated haemolysis.

Treatment [edit]

The treatment for Evan's syndrome will involve close monitoring of hemoglobin and platelet counts. To increase the number of platelets and RBCs in the blood, the patient may be given a corticosteroid such as prednisone. Prednisone will reduce the destruction of the platelets and the RBCs. A certain level of prednisone may need to be taken for the patient's entire life to prevent the levels of platelets and RBCs from dropping. Further treatment may be necessary with immune system suppressing drugs to block the

production of the antibodies. To specifically treat the anemia of the syndrome, it may be necessary to provide the child with new RBCs by a blood transfusion. It has not been shown to be effective to transfuse platelets in a similar fashion to treat the thrombocytopenia. In more severe cases that are unresponsive to treatment, it may become necessary to remove the spleen. The spleen is the organ that is responsible for removing the platelets and RBCs from circulation in the blood once they have been marked with the antibodies. A person can live a normal life without his spleen but needs to avoid injury as much as possible.

Prognosis [edit]

Given the fact that the signs and symptoms of this disease are related directly to the abnormal laboratory findings, the prognosis depends on the patient's response to treatment. Spontaneous remissions of each of the individual component conditions have been reported. If the child responds well to the treatment and the levels of platelets and red blood cells increase, the child can expect to live a normal life. Medications will be needed life long, and laboratory tests will need to be constantly monitored to detect any abnormal changes so that treatment can be adjusted.

Evan's Syndrome is rare, serious, and has a reported mortality rate of just under 18%.

The future [edit]

In a patient with refractory disease, an allogeneic hematopoietic stem cell transplant (HSCT) resulted in complete clinical and serologic remission for more than 30 months. Allogeneic HSCT may be the only current curative therapy for Evans syndrome, but may also be complicated by significant toxicities.

External links [edit]

- http://www.evanssyndrome.net

References [edit]

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