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Case Report

A case of Evans syndrome

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ABSTRACT

Evans syndrome is an autoimmune disorder described by Robert Evans in 1951 specifying that a link exists between primary thrombocytopenic purpura and acquired haemolytic anaemia. It is a rare autoimmune disorder characterised by the simultaneous or sequential development of autoimmune haemolytic anaemia (AIHA) and immune thrombocytopenia (ITP) with or without immune neutropenia. Approximately, Evans syndrome represent 5-10% of warm autoimmune haemolytic anaemia cases and 2-5% of ITP cases. It is basically a diagnosis of exclusion. We present a 50-year-old female presented with anaemia, jaundice and petechia. Patient indirect bilirubin was raised, Coombs test was positive, lactate dehydrogenase and reticulocyte count were raised. Based on the Coombs positive haemolytic anaemia and thrombocytopenia patient was diagnosed with Evans syndrome. ANA profile was positive for SS-A suggestive of underlying autoimmune disorder. Evans syndrome is difficult to diagnose and treat. Typically, corticosteroids and other immunosuppressive medications are used to manage the syndrome.

Keywords: Evans syndrome, Autoimmune hemolytic anemia, Idiopathic thrombocytopenia

INTRODUCTION

Evans syndrome is an autoimmune disorder described by Robert Evans in 1951 specifying that a link exists between primary thrombocytopenic purpura and acquired haemolytic anaemia.[1] It is a rare autoimmune disorder characterised by the simultaneous or sequential development of autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia (ITP) with or without immune neutropenia. Approximately, Evans syndrome represent 5-10% of warm autoimmune hemolytic anemia cases and 2-5% of ITP cases.[2] It is basically a diagnosis of exclusion.

CASE REPORT

A 50-year-old female presented with chief complaints of shortness of breath and generalised weakness since 6 weeks, yellowish discolouration of eyes since 3 weeks, not associated with purities and high coloured urine. No history of similar complaints in the past. Born out of non-consanguineous marriage. Married and having 2 children. On examination she is Afebrile, anaemic, icterus present and petechia was present. Her vitals were stable. On systemic examination mild splenomegaly was present. Laboratory examination results revealed haemoglobin 4.7 g/dl, mean corpuscular volume (MCV) - 97 f/l, mean corpuscular hemoglobin (MCH) - 29 pg, mean corpuscular hemoglobin concentration (MCHC) - 33 g/dl, white blood cell (WBC) - 11,400 cell/cumm, and platelet - 35,000/cumm. Peripheral blood smear showed mild anisopoikilocytosis comprising normocytes and macrocytes and few spherocytes. Reticulocyte count was 2.5% and lactate dehydrogenase (LDH) was elevated. Erythrocyte sedimentation rate was 140 mm at the end of first hour. Direct and indirect Coombs test was positive. Her renal function tests were normal. Serology test was non-reactive.

Liver function tests showed total bilirubin of 6.7 mg/dl and in direct bilirubin of 4.9 mg/dl. Her prothrombin time, activated partial thromboplastin time and international normalised ratio results were normal. Ultrasonography (USG) abdomen was done showed mild splenomegaly. 2D echocardiography (Echo) and chest X-ray showed normal study. Bone marrow examination was done and it showed megakaryocytes and normoblastic erythropoiesis. Based on the Coombs positive haemolytic anaemia and thrombocytopenia patient was diagnosed with Evans syndrome. ANA immunofluorescence tested positive. ANA profile was positive for SS-A suggestive of underlying autoimmune disorder. Then patient was diagnosed with secondary Evans syndrome associated with Sjögren's syndrome. Initially 2 units of packed red blood cells were transfused. Then patient was treated with methyl prednisolone 500 mg intravenously for 3 days, then started on gradually tapering dose of oral prednisolone starting from 1 mg/kg/day. Laboratory reports after 10 days showed haemoglobin of 8.4 mg/dl, platelets of 86,000/cumm and total bilirubin of 2.7 mg/dl. Her symptoms were improved and she responded dramatically.

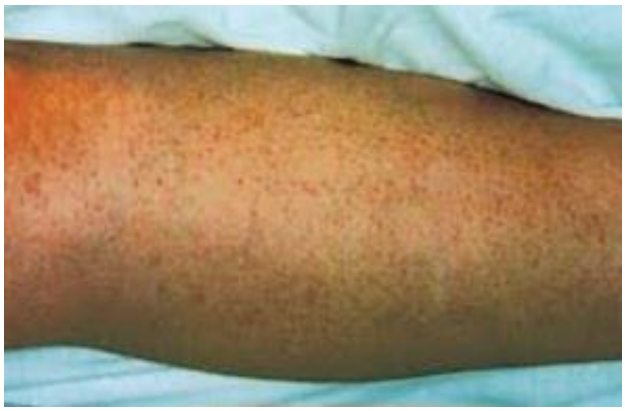

Figure 1: Petechia.

DISCUSSION

Robert Evans originally described the Evans syndrome in 1951. The diagnosis is uncommon and necessitates a high degree of suspicion in order to rule out other conditions that exhibit autoimmune haemolytic anaemia and thrombocytopenia. The cause of the condition is unknown, but immunological dysregulation may play a role in its pathogenesis. Although these defects of the immune response are present in other auto-immune illnesses, constitutive synthesis of IL-10 and INF may activate autoreactive, antibody-producing B cells.[3] Recent studies emphasise that immunisation may act as a catalyst for the onset of disease in susceptible people.[4]

Evans syndrome can present with either thrombocytopenia-related symptoms (purpura, petechiae, ecchymoses, mucosal bleeds in the form of menorrhagia, haematuria, and gastric haemorrhage), or anaemia-related symptoms (pallor, fatigue, and light-headedness) or symptoms of both.[5] Physical examination may also reveal lymphadenopathy, splenomegaly. The diagnosis of haemolytic anaemia requires direct agglutination test (DAT) positivity, peripheral smear for presence of spherocytes can provide variable clues in addition to laboratory findings of elevated lactate dehydrogenase, reticulocytosis and elevated indirect bilirubin.[6] It necessary to exclude infiltrative process in patients with pancytopenia, especially before starting corticosteroid therapy.

Evans syndrome can be primary (or idiopathic) or secondary associated with autoimmune disorders like systemic lupus erythematous, Sjögren's syndrome, anti-phospholipid syndrome and autoimmune lymphoproliferative syndrome. In a study done in 2009, Evans syndrome was found to be "primary" in 34 individuals (50%) but was linked to an underlying condition in 50% of cases, primarily systemic lupus, lymphoproliferative diseases, and common variable immunodeficiency.[7]

The management of Evans syndrome remains a challenge. The syndrome is characterised by periods of remission and exacerbation. Most patients react to corticosteroids and/or intravenous immunoglobulin as first-line treatments, but relapses are common. Steroids are given at 1 to 2 mg/kg per day tapered over weeks in isolated ITP or over months when warm AIHA is present. Immunosuppressive medications, particularly ciclosporin or mycophenolate mofetil, vincristine, danazol, or a combination of these medications are available as options for second-line therapy.[8] Rituximab or splenectomy may be considered in those refractories to the standard treatment or if steroid-dependent that is, at least prednisone greater than or equal to 15 mg required daily to prevent relapse.

CONCLUSION

Evans syndrome is typically characterised by the simultaneous or sequential occurrence of two or more cytopenias. The two that occur most frequently are ITP purpura and autoimmune haemolytic anaemia. Evans syndrome is difficult to diagnose and treat. Typically, corticosteroids and other immunosuppressive medications are used to manage the syndrome. The response, nevertheless, is unpredictable and inconsistent. Evans syndrome patient's prognosis are guarded; those who respond well to treatment get positive results.

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