A Rare Case of Evan Syndrome with Portal Hypertension

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Abstract: Evan's syndrome was first described in 1951 by Evan's and associates. It has long been considered as a coincidental combination of ITP and AIHA and or immune neutropenia in the absence of any underlying cause. We report this rare condition in a 9-year-old male who presented with severe pallor and multiple ecchymotic patches over arms and abdomen with portal hypertension.

Keywords: Evans, Autoimmune, Thrombocytopenia.

INTRODUCTION

Evan's syndrome is an autoimmune disorder characterized by simultaneous or sequential development of autoimmune haemolytic Anaemia (AIHA) and idiopathic thrombocytopenia purpura (ITP) [1-3]. Evan's syndrome is a rare disorder because it is diagnosed in only 0.8% to 3.7% of all patients with either ITP or AIHA at onset. Evan's syndrome was first described in 1951 by Evan's and associates. We report this rare condition in a 9-year-old male who presented with severe pallor and multiple ecchymotic patches over arms and abdomen with portal hypertension.

CASE REPORT

9-yr-old male child presented with 7-day history of increasing lassitude,weakness, palpitation and fever. One day prior to admission he developed haematemesis and malena. There is no relevant past or family history of similar illness. There is no history of any drug intake or toxin exposure. There is no history of any blood transfusion.

The physical examination revealed anxious child with pulse rate 100/min, Respiratory rate 36/min, Temperature of 100*fahrenheit, capillary refilling time less than 2 sec, Blood pressure of 110/70mm Hg, marked pallor with mild icterus. The weight was 28kg (50^{th} centile) and height was 137cm (75^{th} centile). There was multiple ecchymotic patches over arms and abdomen. There was angular stomatitis with mild cheilitis. There was no significant lymphadenopathy. The systemic examination revealed liver span of 12cm with spleen 2cm below left subcostal margin. The other system was essentially normal.

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The laboratory parameters revealed hemoglobin 4.4gm%, hematocrit 13.2%, MCV 88fl, MCH 28pg, MCHC 32gm/dl, total leucocytic count 12000/cmm with normal differential count. The platelet count was 50,000/cmm. The total serum bilirubin was 3.2mg% with SGOT/SGPT/ALP 52/21/98 IU/L, the serum LDH was 3640IU/L(N upto 450IU/L). The renal function test and electrolytes were normal. The peripheral blood smear showed marked aniso poikilocytosis with corrected retic count of 12% with fragmented cells suggestive of severe intravascular hemolysis. The direct coombs test was positive and anti platelet antibody was positive. The BT, CT, PT and APTT were within normal range. His VDRL, HBsAg, HIV, ANA was negative. The bone marrow examination revealed erythroid hyperplasia with megakaryocyte with no leukemic cells. The ultrasonography and Doppler flow study of the abdomen and portal vein showed hepatosplenomegaly with normal echotexture of liver with raised portal venous pressure. The chest CT scan was normal and abdomen CT scan with Doppler flow revealed hepatosplenomegaly with increased portal venous pressure. The UGI endoscopy was done which showed grade 1 varices. Based on the above findings the diagnosis of autoimmune haemolytic anaemia with idiopathic thrombocytopenic purpura (Evans Syndrome) with portal hypertension was made. The child was started on prednisolone in the dose of 2mg/kg/day and propanolol in the dose of 1mg/kg/day. The child responded to the treatment within two weeks. His haemoglobin increased to 6gm% and platelet count increased to 1lac/cmm and corrected reticulocyte count decreased to 5%. He is under follow up with us.

DISCUSSION

Evan's syndrome was first described in 1951 by Evan's and associates. It has long been considered as

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a coincidental combination of ITP and AIHA and or immune neutropenia in the absence of any underlying cause. More recently the spectrum of the disease has broadened specially in children and there is increasing evidence to suggest that Evan's reflects the state of profound immune dysregulation as opposed to coincidental combination of immune cytopenias. Evan's Syndrome can be classified as primary (Idiopathic) or secondary (Associated with some disease). There is increasing evidence to suggest that ES may be associated with or show other diseases or conditions such as systemic lupus erythematous (SLE) [4], lymphoproliferative disorders [5, 6]. The diagnosis of ES requires the simultaneous presence of coombs' positive hemolytic anemia and thrombocytopenia without any other obvious cause. An upper limit of 10 years has been proposed by some to prevent a diagnostic bias. The role of anti-platelet antibodies is yet under review. Other causes of bicytopenia including thrombotic thrombocytopenic anemia or a lymphoproliferative disorders need to be excluded carefully. With the recognition of autoimmune diseases and lymphoproilferative diseases that may be underlying in many cases, a workup including chest and abdominal tomography, ANA studies, serum protein, and immunoglobulin electrophoresis have been proposed as part of the initial workup. Although steroids remain the first-line of management and despite a good initial response (as high as 80% in most series), sustained response rates are lower, and second-line agents like danazol, dapsone, aza-thioprine, cyclophosphamide, mycophenolatemofetil, cyclosporine have been used with varying degrees of success and need to be considered early. Splenectomy is an established form of second-line therapy, but responses are poorer than in AIHA or ITP individually. Rituximab has been tried and found to be at least as successful as splenectomy [7, 8].

In our case Evan syndrome was diagnosed based of autoimmune haemolytic anaemia with idiopathic thrombocytopenia. The exact cause of portal hypertension in this case could not be deciphered. This case is associated with portal hypertension which has never been reported in the literature.

CONFLICT OF INTEREST

None.

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