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Case Report

A Case of Evans Syndrome in Autoimmune Haemolytic anaemia patient with Transfusion related acute lung injury

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ABSTRACT:

Evans syndrome is an autoimmune disease in which an individual's antibodies attack their own red blood cells and platelets. Both of these events may occur simultaneously or one may follow on from the other. Its overall pathology resembles a combination of autoimmune haemolytic anemia and immune thrombocytopenic purpura. There are very few cases reported of Evans syndrome. Here, we present a rare case of transfusion related acute lung injury suspected to be Evans syndrome.

Key words: Evan syndrome, Haemolytic anaemia, immune thrombocytopenic purpura.

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INTRODUCTION

Evans syndrome is an autoimmune disease in which an individual's antibodies attack their own red blood cells and platelets. Both of these events may occur simultaneously or one may follow on from the other. Its overall pathology resembles a combination of autoimmune hemolytic anemia and immune thrombocytopenic purpura.[1-3] [Fig 1]

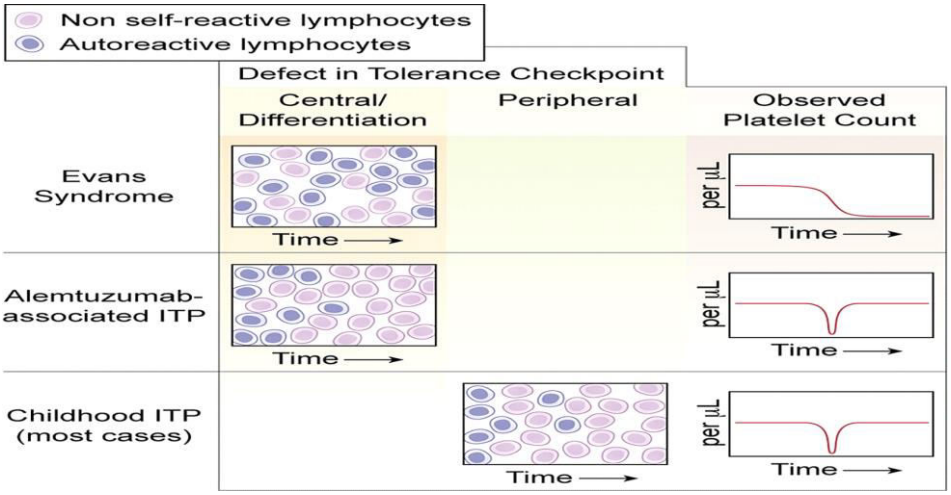

Fig 1: Relation between the autoimmune reaction and thrombocytopenia in Evans Syndrome.

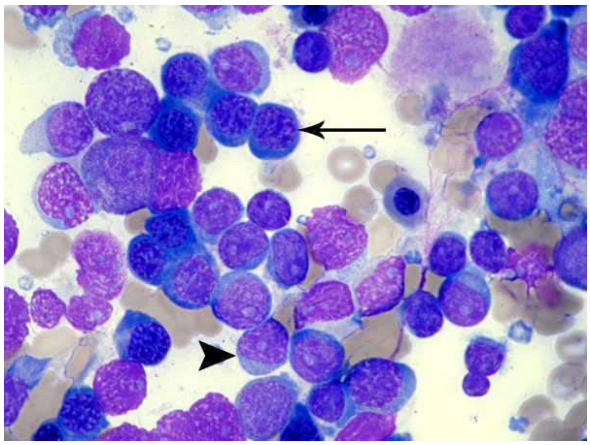

Fig 2: Peripheral smear in Evans Syndrome post autoimmune reaction.

CASE STUDY:

A 37 years old female reported to the casualty with chief complaint of breathlessness from last 4 months (subsided on rest, grade 3 NYHA aggravated more in the last 4 days), palpitations from last 3 days, low grade fever since 3 days (continuous. subsided on taking medications, associated with headache) and vomiting since once day (6-7 episodes, containing food particles, non-bile, non-projectile, non-blood stained). She had no history of chest pain, syncope, any bleeding 0tendencies, pain in abdomen, loose stools, constipation, loss of, trauma or seizures. She had no medical history of diabetes mellitus, hypertension, Koch' s or Koch's contact. Her bowel / bladder habits were normal, and she had achieved menopause about one year ago. She did not have any significant obstetric history, no history of any addictions, and no significant family history. She underwent cataract surgery (right eye) 3 months ago. On examination, it was revealed that she has petechiae over the limbs with pallor ++, pulse was 140 beats per minute, blood pressure was 100/70 mmHg and Spo2 was 96%. On systemic examination, CVS revealed S1 and S2 with no murmurs, respiratory system revealed air entry bilaterally equal with no adventitious sounds heard, PA was soft with no G/T/R, and she was conscious and oriented. Patient was admitted in MICU on March 26, 2017 at 12:55 am.

Following are her blood work reports on March 25, 2017

- Hb-2.2
- S. creatine 0.5
- Retic count – 4.8%
- TLC -16.2
- T.B 2.2 (1.2/1.0)
- LDH - 621
- Platelets count -3.0
- T.P- 5.0 (albumin – 3.2)
- S. Iron – 17.5
- PCV -9
- SGOT/SGPT - 26/23
- MCV -70
- ALP – 98

Peripheral Smear: Severe microcytic hypochromic with anisopoikilocytosis, pencil cells, ring cells, 2 pint PCV transfusion was given.

On March 26, 2017, her overall examination revealed moderate GC, afebrile, petechiae over the limbs with pallor ++, pulse was 130 beats per min, blood pressure was 100/70 mmHg, and Spo2 was 98%. Systemic exam revealed CVS-S1 S2 + with no murmurs heard, respiratory system examination revealed air entry bilaterally equal, and no adventitious sound heard.

Post transfusion blood work reports:

- Hb-6.6,
- TLC-13.2,
- Plt-10.0,
- PCV-20,
- MCV-81
- 1pint PCV transfusion, 1pint SDP transfusion was given.

On March 27, 2017, provisional diagnosis was established as autoimmune haemolytic anaemia. GC was moderate, afebrile, petechiae over the limbs with pallor ++, pulse was 116 beats per min, blood pressure was 110/70 mmHg, and Spo2 was 98%.

Systemic exam revealed normal CVS, and respiratory systems.

Blood work reports:

- Hb-4.6,
- TLC-12.4,
- PLT-8.0,
- PCV-15,
- MCV-81

Repeat blood work"

- HB – 4.6,
- TLC– 6.1,
- PLT– 3.0,
- PCV-15,
- MCV-81

Direct Coombs test: POSITIVE

On March 27, 2017 at 7:30 p.m., there was sudden onset of breathlessness, patient was agitated.

General exam revealed poor general condition, pulse at 150 beats/min, blood pressure at 50/100 mmHg, and respiratory rate at 38/min, Spo2 was 94% on Fm @ 6lts/min. CVS exam revealed S1 and S2 +, and no murmurs heard. Respiratory exam revealed reduced air entry bilaterally with bilateral basal crepitus. CNS was consious and agitated. Blood transfusion was stopped. Inj Lasix 40 iv stat was administered, InjAvil1 amp iv stat, InjHydrocort 1amp iv stat.

On March 27, 2017 at 9:30 p.m., patient was intubated because of dropped Spo2 at 70% on 02 via FM @ 6lts/min. patient was at gasping state, and unconscious.

PRVC: 100/5/26/320

At 12:00 a.m., on March 28, 2017, there was sudden fall in blood pressure (50 mmhg). A Central line insertion was done. InjNorad started at 3.0 ml/hr. At 3:00 a.m., the output also tanked and reduced to 5ml/hr. The BP of the patient was not recordable more than 50mmhg even after starting on double ionotropes, Spo2- 60% on Prvc (100/5/26/320). At 8:00 a.m., provisional diagnosis was established as auto immune haemolytic anaemia with septicaeimia. General condition was poor,febrile, petechiae over the limbs, and pallor ++. Pulse was at 56 beats/min, blood pressure at 50 systolic mmHg, Spo2 at 60% on (PRVC 100/5/26/320). CVS exam revealed S1 and S1 + with no murmurs heard. Respiratory system exam revealed decreased air entry decreased bilaterally lower zone, bilateral crepts positive, CNS was sedated and paralysed, pupils were fixed and dilated. CPCR started and ionotrope support increased.
At 9:00 a.m. on March 28, 2017, patient could not be revived even after all the resuscitation measures. Patient was declared dead on March 28, 2017 at 9:15 am. Cause of death being: Transfusion related acute lung inury in a c/o Autoimmune Haemolytic anaemia suspected to be Evans Syndrome.

DISCUSSION:

Anamika et al reported a case of TRALI in a patient who underwent laparotomy for ruptured corpus luteal cyst requiring blood transfusion. She presented with acute pulmonary edema about an hour after commencing a blood transfusion. This was managed conservatively with oxygen, steroids and diuretics. Patient improved rapidly and later discharged without any residual complications. Lin Y et al described a case of transfusion-related acute lung injury (TRALI) after platelet transfusion immediately following cardiac surgery. A 62-yr-old man was transferred to centre for urgent coronary artery bypass grafting in the setting of recent anti-platelet medication use. Soon after surgery he received platelet transfusions despite having only moderate blood loss. Shortly following the platelet transfusion, he suffered acute hypoxic and hypotensive decompensation requiring nitric oxide therapy, inotropic support, and prolonged need for mechanical ventilation. The patient was eventually discharged from the intensive care unit nine days following the event. The diagnosis of TRALI was made by clinical and radiographic criteria. Donelan KJ et al reported a 3-year-old male with a history of acute lymphoblastic leukemia (ALL) developed TRALI after receiving three units of platelets and a partial unit of packed red cells. He recovered after 24 hours in the pediatric intensive care unit. Laboratory investigation revealed that two of the four blood donors, from which the platelets and packed red cells had derived, had positive human leukocyte antigen (HLA) antibody screens. Further testing of these two donors revealed that one had a specific HLA antibody matching an antigen of the patient. This donor was implicated in the TRALI reaction. TRALI is often mistaken for other transfusion reactions, most notably pulmonary edema caused by circulatory overload or congestive heart failure. It is difficult to gauge which transfusion recipients are at risk for TRALI. Good judgment and transfusion practices when ordering blood products and recognition of the clinical manifestations, diagnosis and treatment of TRALI is critical.[4-6]

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