Intracranial Haemorrhage as a Fatal Complication of Evans Syndrome: A Case Report

Olita Shilpakar,1 Bibek Rajbhandari,2 Bipin Karki,3 Umesh Bogati1

1 Department of General Practice and Emergency Medicine, NAMS, Bir Hospital, Kathmandu, Nepal
2 Department of General Practice and Emergency Medicine, Nepal Police Hospital, Kathmandu, Nepal
3 Department of Critical Care Medicine, Om Hospital and Research Centre, Kathmandu, Nepal

ABSTRACT

Evans syndrome is a rare hematologic disorder characterized by the presence of simultaneous or sequential direct Coombs-positive autoimmune hemolytic anemia (AIHA), immune-mediated thrombocytopenia and/or immune neutropenia without any known underlying etiology. Spontaneous intracranial hemorrhage is a rare and life-threatening complication in patients with Evans syndrome and very few cases have been reported to date. We report a case of a thirty-two- year-old female with intracranial haemorrhage with underlying Evans syndrome who presented with the clinical manifestations of headache, vomiting and altered sensorium and succumbed to the fatal complication despite resuscitative measures. This also emphasizes the importance of early recognition of symptoms and immediate presentation to health care facilities for aggressive management of the patient.

Keywords: autoimmune; evans syndrome; intracranial haemorrhage.

INTRODUCTION

Evans syndrome is an uncommon hematologic condition comprising anemia and thrombocytopenia.1 There is no preferential distribution by age, sex or ethnicity and its course is characterized by recurrent exacerbations and remissions.1 Patients are at risk of bleeding manifestations and may include rare but life threatening complications like intracranial haemorrhage due to severe thrombocytopenia.1,2 Therapy includes corticosteroids, intravenous immunoglobulins, immunosuppressive agents and splenectomy.1,2

CASE REPORT

A thirty-two-year-old female presented to the Emergency Room (ER) with the history of generalized headache since the past three days not relieved by paracetamol taken from a local medical shop. The severity of headache increased in intensity and by the end of day two she started having multiple episodes of non bilious, non projectile vomiting. She was finally brought to the ER after she became unresponsive. The patient's family denied any history of fever, seizures or trauma. She was a diagnosed case of Evans Syndrome with chronic idiopathic thrombocytopenia since the past five years and was under steroid therapy and an immunomodulating agent azathioprine, however, she had not been compliant with her medications since the past three weeks. She had regular menstrual cycles with history of one spontaneous abortion around seven years back.

On examination in the ER, the patient was unconscious with a Glasgow coma scale score of 5/15 (E1V2M2). Her radial pulse was 125 beats per minute, blood pressure 130/80mm Hg, respiratory rate 28 breaths /minute, temperature 98.2 degrees F and 80% oxygen saturation in room air. Bilateral pupils were fixed with the right and the left pupils 5mm and 3mm in size respectively. Systemic examination did not reveal anything abnormal except bilateral mute plantars. Taking into consideration of her deteriorating condition, immediate resuscitation was started by securing her airway via intubation.

Correspondance:
Dr. Olita Shilpakar,
Department of General Practice and Emergency Medicine, NAMS, Bir Hospital, Kathmandu.
E-mail: olitashilpakar@yahoo.com,
Phone: 977-9841256959.
with an endotracheal tube of 7 mm internal diameter. A complete blood count showed a hemoglobin of 7 gm% and severe thrombocytopenia with a platelet count of 8,000/mm³. Renal and hepatic functions including a coagulation profile were unremarkable. A plain computer tomography (CT) scan of the head revealed right parieto occipital intracranial haemorrhage (ICH) with intraventricular extension with perilesional edema and midline shift. (Figure 1) An ICH score of 4 was calculated (GCS: +2 points, age<80 years: 0 point, ICH volume >30ml: +1 point, intraventricular haemorrhage: +1 point, infratentorial haemorrhage: 0 point). The 30-day mortality of the patient as per the ICH score was 97%. Administration of 200ml of 20% mannitol intravenously along with initiation of anticonvulsant therapy was done via a loading dose of 1 gram of levetiracitam. Four pints of platelet rich plasma were asked to be arranged immediately for transfusion. Stabilisation of the patient under strict hemodynamic monitoring in the intensive care unit with mechanical ventilation was planned. However, the patient’s family deferred all possible forms of treatment owing to the poor prognosis and the high ICH score. They agreed not to further continue any resuscitative measures and the patient died a few hours later.

Figure 1. Plain CT head showing right parieto occipital intracranial haemorrhage with intraventricular extension with perilesional edema and midline shift.

DISCUSSION

Evans syndrome is a rare clinical entity initially described by Robert Evans in 1951.² It is considered as a diagnosis of exclusion. The exact etiology and pathophysiology still remain unknown, however, immune dysregulation may be involved in the pathogenesis of the disease. It is associated with non cross-reacting autoantibodies directed against antigens specific to RBCs, platelets or neutrophils.¹² Evans Syndrome can be classified as primary (idiopathic) or secondary which is associated with diseases like systemic lupus erythematosus, primary antiphospholipid syndrome, Sjogren’s syndrome, common variable immunodeficiency and malignant lymphomas.¹² Clinically, symptoms of hemolysis include pallor, fatigue, lightheadedness and jaundice.
Signs of thrombocytopenia include petechiae, bruising, hepatomegaly, splenomegaly, mucosal bleeding and rarely intracranial bleed. In a study of sixty eight patients with Evans Syndrome by Michael et al, 23.5% cases died due to different complications of this disease like septic shock, associated carcinomas, pneumonia, stroke, meningitis, acute myocardial infarction or refractory anaemia. The study inferred that severe thrombocytopenia in Evans syndrome led to two deaths resulting from acute gastrointestinal bleeding and one death from acute intracranial haemorrhage which was similar to our case. Rashid et al also reported a similar case of bleeding manifestation in the form of non traumatic non aneurysmal subarachnoid haemorrhage in a patient with Evans syndrome.

Intracranial haemorrhage is one of the rare but serious side effects of severe thrombocytopenia in patients with hematological disorders which can lead to devastating consequences. The ICH Score is a simple clinical grading scale comprising of five factors independently associated with the 30-day mortality rates like the Glasgow Coma Scale score, age >/=80 years, infratentorial origin of ICH, ICH volume and the presence of intraventricular hemorrhage. This score allows risk stratification and provides information regarding the degree of severity of ICH that helps in making decisions regarding the goals of care. The family's wishes and the patient's comorbid conditions should be taken into consideration like in our case where she had an ICH score of 4 with an estimation of 97% mortality rate.

The management of Evans syndrome remains a challenge to date since the response to various treatment modalities may vary as per the disease progression. Stabilisation of respiratory and cardiovascular functions and blood and blood product transfusion like platelet rich plasma and platelet concentrate aids in alleviation of the symptoms. A platelet count threshold of 50 x 10^9 /L prior to major surgery and 100 x 10^9 /L prior to surgery involving the brain or eyes is recommended.

The first-line of definitive management consists of steroids although frequent relapses following its weaning have been encountered. Other modalities of management include pharmacological therapy with intravenous immunoglobulins (IVIG) and immunosuppressants like vincristine, cyclophosphamide, azathioprine, danazol, mycofenolate mofetil, newer drugs like rituximab and surgical intervention like splenectomy. Our patient was on steroids and azathioprine as the immunomodulator but she was non compliant and had not been taking her medications since the past three weeks which could have led to exacerbations and fatal complications.

Evans syndrome is a relapsing disease which may present acutely with life threatening complications. Our case report highlights the need to acquaint the treating physician regarding rare fatal complications like intracranial haemorrhage with thrombocytopenia. We would also like to emphasize the importance of patient education on compliance, regular follow up, remissions and exacerbations. Newer modalities like Rituximab, a chimeric monoclonal antibody against CD20 and autologous and allogenic stem cell transplantation in refractory cases could be considered to prevent life threatening complications.

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CONSENT: NMJ Case Report Consent Form was signed by the patient's family and the original is attached with the patient's chart.

CONFLICT OF INTEREST: None.

REFERENCES


