



## A RARE CASE OF DELAYED SECONDARY IDIOPATHIC THROMBOCYTOPENIC PURPURA IN A PATIENT WITH TREATED GUILLAIN BARRE SYNDROME.


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### ABSTRACT

The Idiopathic Thrombocytopenic purpura (ITP) associated with concurrent GuillainBarre Syndrome (GBS) has been reported previously but delayed onset ITP following GB syndrome is a rare phenomenon. This is a case of 66 years old male a recovered case of Guillain-barre Syndrome readmitted to our hospital with complaints of hemoptysis. Routine blood investigations were done which revealed Pancytopenia. Bone marrow aspiration and biopsy showed hypercellular marrow and increased megakaryocytes. Patient received multiple platelet transfusions in view of hemoptysis and hematuria. He was diagnosed to be having severe secondary ITP in view of persistent low platelets and hence received steroids, Intravenous immunoglobulin and Eltrombopag (Thrombopoietin receptor agonist). Due to refractory thrombocytopenia patient underwent splenectomy following which platelets started showing an increasing trend within 48 hours.

**Key words:** GuillainBarre Syndrome, Idiopathic Thrombocytopenic purpura, Eltrombopag, Splenectomy.

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### INTRODUCTION

GB syndrome encompasses an acute inflammatory immune-mediated demyelinating polyneuropathy. It occurs by a mechanism called molecular mimicry. ITP is a condition wherein auto antibodies bind to the platelet membrane glycoprotein resulting in sequestration by mononuclear macrophages in the spleen causing variably low platelet counts. Autoimmune conditions often appear to occur concurrently, and ITP often occurs with other autoimmune diseases [1], however, GBS typically does not occur with other autoimmune diseases [2].

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### CASE REPORT

This is a case of 66 years old male, known case of Type 2. Diabetes Mellitus on regular treatment got admitted to our hospital with complaints of hemoptysis and petechiae. He is also a recovered case of Guillain-barre Syndrome (AMSAN type) which was diagnosed 7 months back and was treated with Plasmapheresis and Intravenous immunoglobulin following which he had good neurological recovery. This admission patient was intubated in the ER in view of hemoptysis and shifted to ICU for further management. Routine blood investigations were done which revealed Hemoglobin-8.5gms/dl, Platelet Count-4,000 cells/cu mm, total leucocyte count-2.4cells/cu mm. The MRI brain was normal. No hepato-splenomegaly. Coagulation parameters (PT/ aPTT) were normal, Serology and ANA profile negative. Peripheral smear study showed

normocytic normochromic anemia. Direct Coombs test was negative. Bone marrow aspiration and biopsy showed hypercellular marrow and increased megakaryocytes. Patient was extubated next day and received multiple units of platelet transfusion for persistent low platelet count of (3000 cells/cumm) and hemoptysis. Patient was diagnosed to be having severe secondary ITP in view of persistent low platelets and hence received prednisone (60mg/day) and Intravenous immunoglobulin 1G/kg. Due to persistent thrombocytopenia complicating as hematuria and hemoptysis, he was started on Eltrombopag which did not show any improvements in platelet count even after 2 weeks of therapy. After multi consensus discussion to overcome the refractory thrombocytopenia, splenectomy was performed under adequate cover of pre splenectomy prophylaxis and blood products. There were no procedure related complications and post-operatively platelets started showing an increasing trend reaching 74,000/cumm on day 2. Later patient was shifted to ward and discharged home in a hemodynamically stable condition with a platelet count of 1.8 lakhs / cumm.

## DISCUSSION

GuillainBarre Syndrome is an immune mediated polyneuropathy which follows an antecedent infection [3]. AMSAN is the severe variant of GBS, which is a B-cell-mediated macrophage, induced demyelinating disease that affects both the ventral and the dorsal horns, thereby causing both motor and sensory deficits. GuillainBarresyndrome (AMSAN variant) with concurrent ITP has been rarely reported [4, 5]. ITP has been defined as immune thrombocytopenic purpura which is an autoimmune disorder where an unknown stimulus leads to autoimmune destruction of normal platelets. Antigenic targets of ITP auto antibodies are GpIIb / IIIa and GpIb / IX [6, 7]. Immune thrombocytopenic purpura has been classified as primary (where cause is not known) or secondary (associated with some disorder which includes infection such as Hepatitis C, HIV, CMV or H pylori. Dugs like thiazide and auto immune disorders like APLA, SLE or Evans syndrome). The antibodies associated with the variants of GBS are directed against gangliosides specifically found in the makeup of myelin [2] Gangliosides are also found in platelets; however, those gangliosides do not correlate with the gangliosides found in the nervous system [8]. Instead, ITP is characterized by an autoimmune reaction against the glycoproteins GP IIa-III b or GP Ib IX. Although the autoimmune reaction is

usually targeting the glycoproteins of platelets in circulation, an autoimmune process also occurs in the bone marrow, causing decreased production of platelets by the megakaryocytes [9]. No overlapping antigen has been discovered between GBS and ITP. Diagnosis is based on thrombocytopenia < 1 lakh/cumm, absence of other causes of thrombocytopenia, absence of anemia, normal leucocytes and increased reticular count but rarely it may present with Pancytopenia like our case. On peripheral smear normocytic normochromic anemia was detected, and bone marrow aspiration showed hypercellular marrow and increased number of megakaryocytes which is consistent with ITP. Treatment is recommended for newly diagnosed patients whose platelet count < 30,000/cumm [10]. First line of treatment is long course of corticosteroids (Prednisone 1-2 mg/kg) [11]. IVIg (1 gm/kg) is recommended when rapid increase in platelet count is required or if corticosteroids are contraindicated [12]. Splenectomy is considered who fail the initial treatment [13]. Thrombopoietin receptor agonists such as Eltrombopag are used who relapse after splenectomy or having contraindication to splenectomy [14]. ITP secondary to GB syndrome is a rare phenomenon and early detection with appropriate treatment can reduce morbidity and mortality. The previously reported cases of GBS and ITP were concurrent [15,16] whereas our patient presented as a delayed case with refractory ITP necessitating splenectomy .This can be attributed to patients developing cross reactivity against both platelet and neurogenic glycoproteins following an antecedent infection and this case emphasizes the treatment decisions related to the finding of delayed ITP in GBS.

## STATEMENT OF HUMAN AND ANIMAL RIGHTS

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors

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## DECLARATION OF INTEREST

None declared.

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