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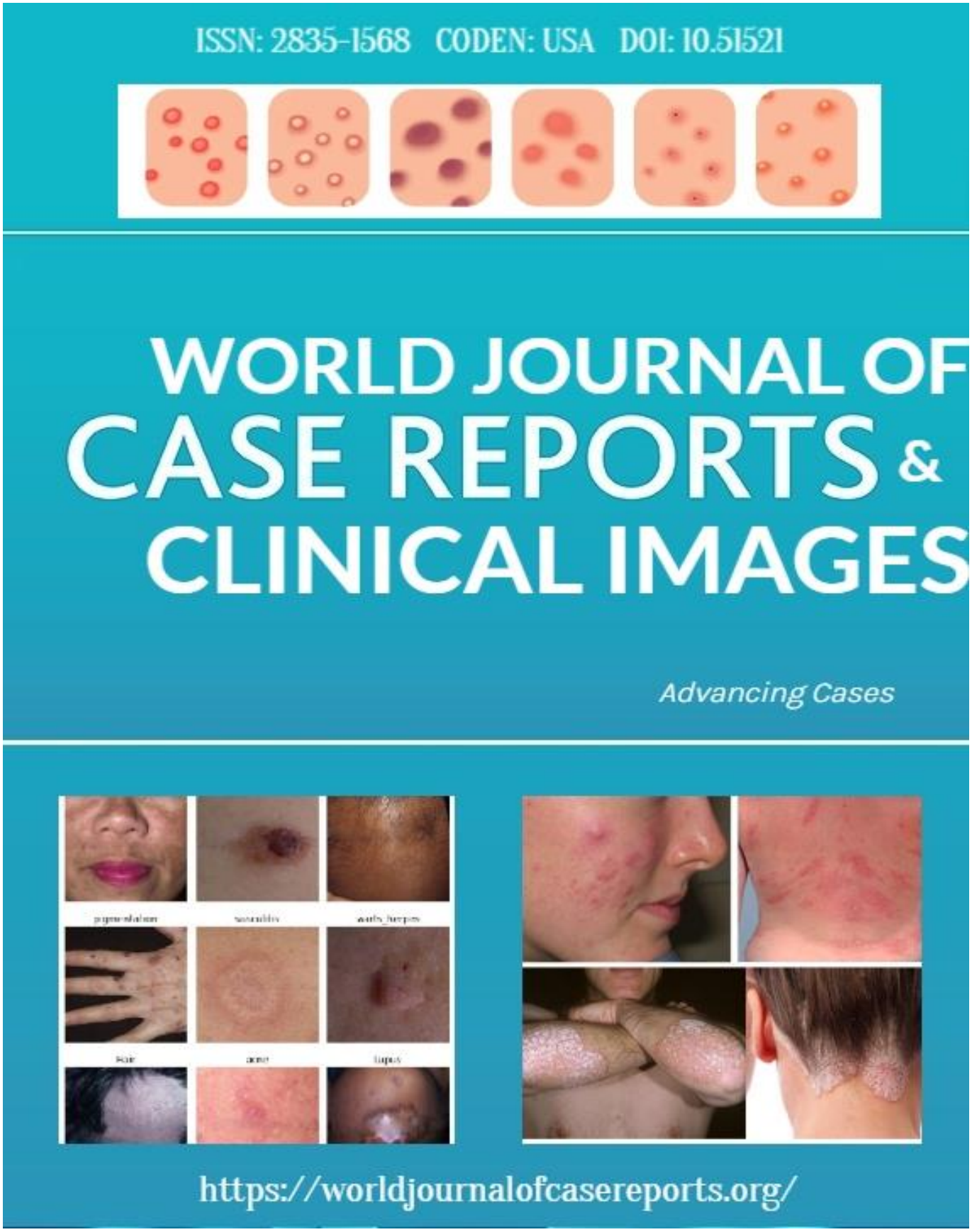

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CASE REPORT

The Utility of Immature Platelet Fraction Monitoring During a Case of Life-Threatening ITP Predicted Haemorrhagic Risk

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Introduction

A severe case of immune thrombocytopenia purpura ITP presented with life threatening bleeding. Despite first line steroids and intravenous immunoglobulin IVIg this patient continued to deteriorate requiring full dose elthrombopag, romiplostim, cyclophosphamide, anti D, mycophenolate mofetil MMF, 31 units of platelets and 14 units of packed red cells. It took 3 weeks before the platelets responded, and the patient went on to make a full functional recovery despite severe pulmonary haemorrhages and a pneumothorax.

Immature platelet fraction IPF measurements are now more readily available given the ability of high-end full blood count analysers to perform them. A high IPF percentage would correlate with increased megakaryopoiesis in the bone marrow. When combined with a thrombocytopenia it would favour a consumptive process like ITP as the underlying aetiology.

In this case IPF was monitored concurrently with the absolute platelet count. Interestingly when the IPF fell during their admission the patient demonstrated overt bleeding. If the bleeding phenotype can be more accurately risk stratified this could help the clinician identify lower risk patients allow them to be managed in an outpatient setting. A larger study of ITP patients is required with IPF measurements to see if a robust risk stratification is possible.

Case Presentation

Mr. M presented to the Direct Assessment Area with bleeding gums and spontaneous bruising. He gave no history of a recent viral illness or any red flag symptoms, and no recent medications including vaccinations were administered.

On examination, he was clinically well and haemodynamically stable. He had a petechial rash over his lower extremities and areas of ecchymosis over his arms and torso.

Blood tests revealed a platelet count of 3 ($x10^9$ /L) with a normal haemoglobin and white cell count. No haemolysis was evident, and haematinics were replete. After morphological examination of the blood film confirmed a true thrombocytopenia and a coagulation screen came back normal the working diagnosis was Immune Thrombocytopenia Purpura ITP.

Further investigations were performed including: a chest Xray; a viral screen- HIV, Hepatitis B and C, Cytomegalovirus, Epstein Barr Virus, H pylori and a CT chest, abdomen, pelvis all of which demonstrated no trigger for the ITP.

He was started on Prednisolone 1mg/kg (80mg) daily. Unfortunately, within several days frank haematuria occurred and as

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the platelets were still low, he received emergency intravenous immunoglobulins (IV Ig) 1g/kg for 2 consecutive days.

On the night after the IV Ig he developed a cough and haemoptysis. At the same time, melaena was evident and progressive ecchymosis. His haemoglobin dropped down to 67. Given this emergency multiple platelet transfusions were administered. Rituximab was infused and elthrombopag was started. A bone marrow biopsy was performed to exclude any other pathology driving the process but later was reported in keeping with ITP.

Mr M continued to deteriorate requiring packed red cells to maintain his haemoglobin and platelets as required. Oxygen requirements were increasing steadily, elthrombopag was increased to maximum dose as well as romiplostin. He reached the maximum oxygen that could be delivered on the ward therefore the risk/benefit approach was taken and cyclophosphamide with vincristine chemotherapy was administered. Interventional radiology was consulted for possible splenic embolisation and surgeons for consideration for an emergency splenectomy.

Mr M entered ICU for further oxygenation with Continuous Positive Airway Pressure CPAP. His oxygen requirements were maintained on CPAP, but an x-ray performed showed a pneumothorax (Image 1). The pneumothorax needed an emergency drain, but as the platelets remained low the haemorrhage risk remained high. During the procedure he received Anti-D as well platelet transfusions and the chest drain was uneventful. In ICU he remained on daily elthrombopag, IV Methylprednisolone, MMF and weekly rituximab and romiplostim. There was no platelet recovery noted for nearly 3 weeks in total. During that time there was no increment with platelet transfusions.

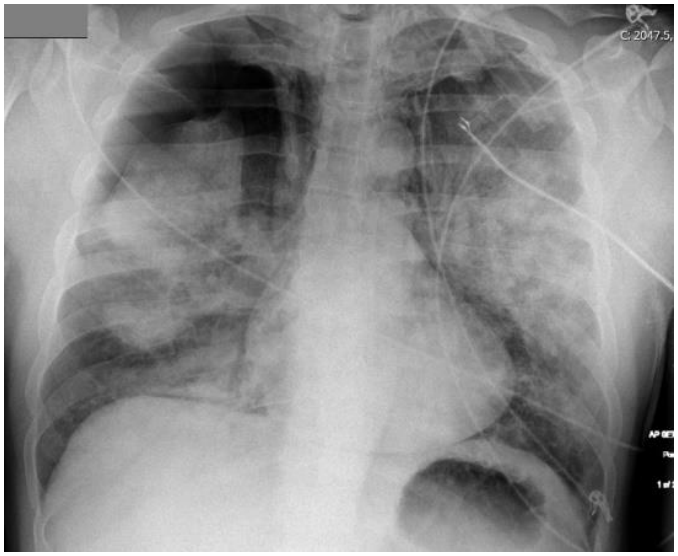
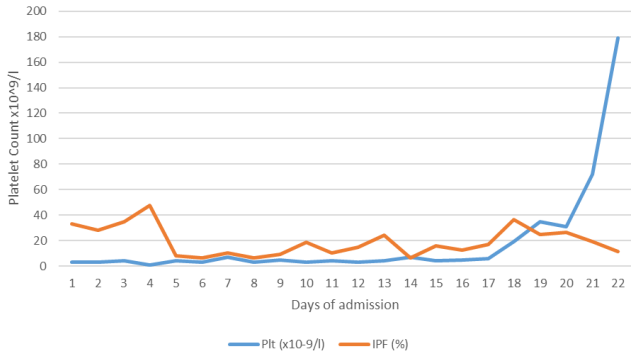


Image 1: Chest X-ray demonstrating extensive pulmonary haemorrhages with a right sided upper pneumothorax.

Mr M started to recover his platelet count around D20 and by D22 his platelets entered normal range. He was initially maintained only on MMF as an outpatient given the severity of his episode. Despite the significant lung injury Mr M made a full functional recovery over the following months. During his hospital admission he received 31 units of platelets and 14 units of packed red cells.

ITP is an acquired autoimmune disorder characterised by thrombocytopenia secondary to increased platelet consumption. Incidence is estimated to be 2-5 per 100,000 in the general population [1]. First line management includes steroids and/or IV Ig, both of which are associated with a good response rate (combined have partial response PR 93% and complete response 78%) [2]. Despite the good response rates this patient failed to respond quick enough and therefore proceeded to second line ITP management with: rituximab (response rates 60%), thrombopoietin receptor agonists TPO-RA romiplostim and elthrombopag (response rates 80-94%) [3,4]. Despite all these treatments with good response rates it took a few weeks before the platelets recovered.

Figure 1-Graph demonstrating the immature platelet fraction IPF% to absolute platelet count during a case of severe ITP until recovery at Day 22.



Recent publications have stated IPF can predict platelet recovery and therefore giving the severity of this case we monitored the IPF daily until recovery (Figure 1). Interestingly the high IPF at presentation correlates with increased megakaryopoiesis in the bone marrow and when associated with thrombocytopenia supports the diagnosis of ITP [5].

From Day 5 to Day 9 the IPF fell to <10% persistently and when this occurred the patient had severe spontaneous haemorrhages of the lungs, bowel, and skin. In severely thrombocytopenic patients the amount of immature platelets may be important as immature platelets are larger and hyper reactive. Therefore, thrombocytopenia with less immature platelets may be more haemorrhagic. Prior to platelet recovery the IPF% increased 24hours before and as the platelets returned to normal range the IPF% fell expectedly.

This case demonstrated despite the good success rates of steroid and IV Ig in the front line setting extreme progressive life-threatening cases can occur. Given the severity of this clinical situation all ITP treatments were administered and eventually the platelet count recovered. Recent literature discusses the utility of immature platelet fraction IPF in the diagnosis and prediction of platelet recovery. This case demonstrated its usefulness for diagnosis but not for predicting recovery in a timely manner or guiding treatment decisions during the acute management. Interestingly this case did demonstrate when the severe thrombocytopenia occurred with an IPF<10% the patient had uncontrollable bleeding. This correlation is useful clinically as thrombocytopenic patients have different bleeding phenotypes. If

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the bleeding phenotype can be more accurately risk stratified this could help the clinician identify a lower risk patient allowing them to be managed in an outpatient setting. A larger study of ITP patients is required with IPF measurements to see if a robust risk stratification is possible.

Conflict of Interest

To the best of our knowledge, no conflict of interest, financial or other, exists with respect to the information provided in this report.

Ethical Consideration: None

Acknowledgements: None

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