Treatment Refractory Immune Thrombocytopenia as a Manifestation of Relapsed Chronic Lymphocytic Leukaemia



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Background

Secondary immune thrombocytopenia (ITP), unlike primary ITP, features the immune destruction of platelets triggered by un underlying immune disorder such as a lymphoproliferative disorder or autoimmune disease.

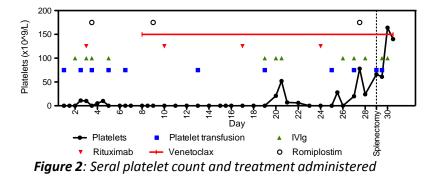


Figure 1: CT Brain demonstrating right occipital intraparenchymal haematoma

1 Cuker A, Neunert CE. How I treat refractory immune thrombocytopenia. Blood (2016) 128(12): 1547-1554 2 Neunert CE et al, American Society of Haematology 2019 Guidelines for immune thrombocytopenia. Blood Adv (2019) 3(23): 3829-3866

Case report

- 79yo male with chronic lymphocytic leukaemia, in remission after chemoimmunotherapy 9 months prior, presented with sudden onset visual loss
- Investigations revealed severe thrombocytopenia (<5 x10⁹/L) and CT brain showed intracranial haemorrhage (Figure 1)
- Of note, peripheral blood lymphocyte count was normal, and there was no appreciable lymphadenopathy or splenomegaly
- Commenced corticosteroids, intravenous immunoglobulin (IVIg) and platelet transfusion support (Figure 2)
- Day 8: persisting thrombocytopenia → rituximab, romiplostim (thrombopoietin receptor agonist) and venetoclax (BCL-2 inhibitor) commenced.
- Day 19: developed gastrointestinal bleeding with no improvement in platelet count → day 29 splenectomy
- 18 days post splenectomy achieved platelet count recovery to 660 x10⁹/L (with ongoing venetoclax, romiplostim and IVIg)



Treatment	Response rate	Time to response
Rituximab	60%	1-8 weeks
Romiplostim	80%	1-4 weeks
Eltrombopag	80%	1-2 weeks
Azathioprine	40-60%	3-6 months
Mycophenolate	11-80%	4-6 weeks
Splenectomy ²	60-70%	1 month

Table 1: Response rates and time to response of common ITP treatments (adapted from Cuker et al¹).

Discussion

- This case highlights the deficiencies of conventional ITP treatments and the importance of treating the underlying disease in secondary ITP.
- Demonstrates the lag time between targeted treatment such as rituximab and romiplostim taking effect (Table 1), which leaves some patients vulnerable to life-threatening bleeding complications despite best medical therapy.

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