Laparoscopic splenic artery ligation in a patient with immune thrombocytopenia with intracranial haemorrhage (two clips that stopped a timebomb)

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SUMMARY

Intracranial haemorrhage (ICH) in a patient with relapse of idiopathic thrombocytopaenic purpura (ITP) can be lethal. The site of haemorrhage, compounded by low platelets in this disease, makes its management extremely challenging, especially when a neurosurgical procedure is warranted. We report a case report of an unconventional way of increasing platelet counts in ITP rapidly in an emergency setting.

KEY WORDS:
Idiopathic thrombocytopaenic purpura; Intracranial haemorrhage; Laparoscopic splenic artery ligation; thrombocytopaenia

INTRODUCTION

Idiopathic thrombocytopaenic purpura (ITP) is a haematological disease characterised by thrombocytopaenia (platelet of less than 100×10^9/L) with unknown causes.[1] The pathogenesis of ITP involves both antibody-mediated and/or T-cell mediated platelet destruction, thus given the name "immune thrombocytopaenia".[2]

Major life-threatening bleeding such as intracranial haemorrhage (ICH) in people with ITP can be disastrous. We report a case of intracranial haemorrhage in an adult with chronic ITP.

CASE REPORT

A 44 -year-old man with three-year history of ITP, who defaulted follow-up and treatment, presented to the Hospital Banting with mild headache after a fall. His Glasgow Coma Scale (GCS) was full and there was no neurological deficit. However, the platelet count of this patient was $3x10^9$/L. Two units of platelets were transfused and a total three grams of intravenous methylprednisolone (high dose steroid) was initiated over a span of three days.

Patient's platelet count ranged from 1 to $3x10^9$/L throughout the three-day stay in the Hospital Banting hospital. He was transferred to Hospital Tengku Ampuan Rahimah (HTAR), Klang on the third day of admission. A Computed Tomography (CT) imaging of the brain was performed and showed left acute on chronic subdural haemorrhage with mass effect, generalised oedema and obstructive hydrocephalus (Figure 1).

The neurosurgeons planned for left burr-hole surgery once the platelet exceeded $90x10^9$/L. The haematology team initiated intravenous immunoglobulin (IVIg) treatment for a total of two grams per kilogram body weight over a span of two days with high dose oral prednisolone one milligram per kilogram body weight per day and four units of platelet transfusion. Despite this, platelet levels continued to remain low.

The general surgical team at HTAR proposed to perform laparoscopic splenic artery ligation. The patient was electively intubated for operation on 11th July 2017. Four units of platelets were transfused before laparoscopic surgery and only one dose of IVIg was given. Platelet count prior to laparoscopic surgery was $25x10^9$/L. The splenic artery was identified at the upper border of pancreas and was clipped at both proximal and distal parts. The artery was not transacted. A further eight units of platelets were prophylactically transfused during the procedure. Blood loss was minimal and there was no postoperative complication. Platelet count immediately post laparoscopic op was $77x10^9$/L. The patient was kept intubated and transferred to a neurosurgical centre.

Twelve hours post splenic artery ligation, his platelet counts increased to 115 and $240x10^9$/L the following day. Left Burr-hole surgery and external ventricular drainage were performed. CT brain postoperatively showed smaller residual subdural haematoma and resolving mass effect (Figure 1).

He was successfully extubated on 16th July 2017 with full GCS and had no neurological deficit. There was no need for further IV Ig and he was put on oral prednisolone. After five days, he was transferred back to HTAR. After splenic artery ligation, he did not require any further platelet transfusion. He was discharged well with immediate doses of standard post-splenectomy vaccines and a tapering dose of oral prednisolone with outpatient follow-up at Haematology Clinic, HTAR.

To date, more than two-year post splenic artery ligation, the patient is well and continues to be in remission. He has resumed work without neurological sequelae.

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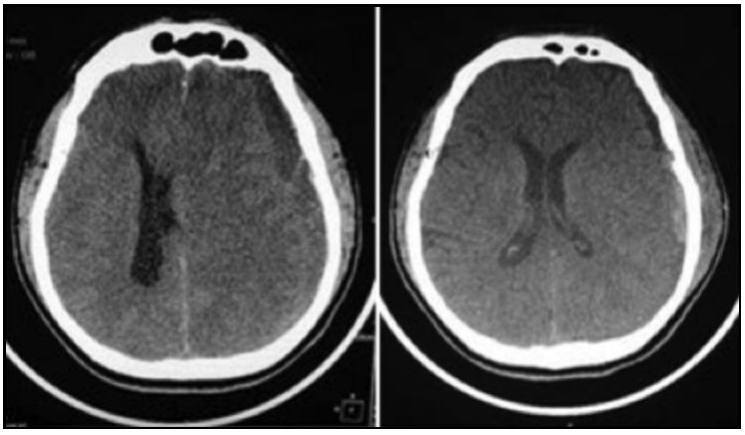

Pre op Post op

Fig. 1: CT brain pre and post operation. The CT brain pre operation showed a left acute on chronic subdural haemorrhage with a midline shift to the right. (left image) After the operation, there was resolution of mass effect and smaller left subdural haemorrhage. (right image).

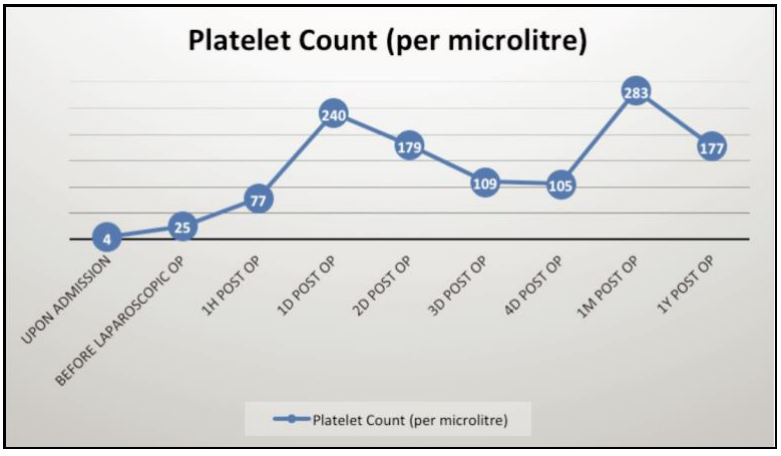

Fig. 2: Trend of platelet counts. The platelet count rose to 77 x 10^9/L an hour after laparoscopic ligation of the splenic artery. The platelet count had remained above 100 x 109/L twenty-four hours after the operation, without necessity of further platelets transfusion or IVIg.

DISCUSSION

Although it is rare, intracranial haemorrhage is the most serious complication in ITP. There are no existing scoring tools or prediction models to determine patients with ITP at risks for ICH. Instead, it is multifactorial, and the risks are different for every patient.

The current treatment approach for all ITP patients with life or organ-threatening bleeding consists of intravenous immune globulin (IVIg) or corticosteroids with platelet transfusion.[1] Pooled normal human immunoglobulin therapy is useful to stimulate rapid platelet increase prior to planned procedures or operations.

This patient suffered from life-threatening intracranial haemorrhage, as evidenced by the significant mass effect with cerebral oedema and obstructive hydrocephalus. An urgent operation to remove the blood clots and relieve intracranial pressure was desired. However, such major operations can never be conducted with platelet counts of $25x10^9$/L. There are no guidelines elaborating on "safe" platelet levels for procedures or operations. The current recommendations for "safe" platelet counts in adults are above $50x10^9$/L for minor surgery and above $80x10^9$/L for major surgery.[2] In this case, there was a need to increase the platelet count urgently.

This man was given adequate first-line emergency medical therapy of high-dose corticosteroids, pooled human immunoglobulin and platelet transfusions, which provided minimal responses.

Rituximab is a chimeric monoclonal antibody directed against CD20, which is a transmembrane glycoprotein expressed on the surface of B-lymphocytes. Upon administration, rituximab induces B-lymphocytes apoptosis or destruction, thus reducing the anti-platelets titres in patients with chronic ITP. However, the median response of platelet rise after rituximab administration was 30 days.[3] We could not to wait that long for a response to be achieved as the patient would have bled further.

Romiplostim, a thrombopoietin (TPO) receptor agonist administered as subcutaneous injection weekly, was approved by the USA Food and Drug Administration in 2008 for adults of chronic ITP who are unresponsive to corticosteroids, IVIg, or splenectomy. It works by activating the TPO receptors, hence increasing megakaryocytes production and is recommended in ITP patients with high bleeding risks. Although its use for haemorrhagic stroke in the setting of ITP was described by Romain Gellens et al., the sustainable platelet recovery was observed only a week after its first administration.[4] For our patient with ICB requiring an urgent surgery, we desired an intervention or treatment method with faster platelet recovery. Another new drug for patients with ITP is eltrombopag, an oral TPO receptor agonist. Similarly, it takes up to two weeks for patients to respond to eltrombopag.

Splenectomy is the second-line treatment approach for patients with chronic ITP who did not respond to corticosteroids, with the complete or partial remission rates achieved in over two-thirds of patients. However, this is a major operation which is almost impossible to be performed in patients with such low platelet count.

In 1973, Maddison reported the clinical experience with splenic arterial embolisation to achieve infarction of splenic tissue. However, it is not deemed a viable option due to significant complications of complete splenic infarction. Since then, partial splenic arterial embolisation (PSE) has been advocated instead.[5]

Kimura and colleagues studied on fifty-one patients with ITP receiving PSE and concluded that PSE is an effective alternative to splenectomy in patients with chronic ITP.[6]

In the setting where interventional radiology services are not available, laparoscopic splenic artery ligation performed by an experienced surgeon is an option. Zhang J.S. and colleagues proposed this technique for the treatment of hypersplenism and thrombocytopenia in children. In their study a total of seven children underwent this surgery from August 2014 till December 2014. They did not receive any transfusion. Post-operative complete blood counts were normal, with evidence of reduction of splenic size.[7]

Till date, there is no case report of this procedure performed in the emergency setting of life-threatening bleeding in patients with chronic ITP.

CONCLUSION

Laparoscopic ligation of the splenic artery is a feasible and safe management option in ITP patients who have major and life-threatening bleeding secondary to thrombocytopenia.

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