Clinical vignette: Zero in 60 in 48 hours

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Background

Cirrhosis is a well known cause of thrombocytopenia, but it is important to consider other etiologies when the degree of thrombocytopenia is severe, especially in cases of life-threatening bleeding.

Case Presentation

34 year-old male presented to the emergency department with hematuria and persistent bleeding from preexisting oral ulcers. He denied melena, hematemesis and hematochezia. He denied any recent change in his medications nor taking any herbal medications or supplements.

Past Medical History:
- Cirrhosis secondary to Hepatitis C and alcohol abuse.
- Pancytopenia secondary to cirrhosis, active hepatitis C infection and hypersplenism.

Pertinent Physical Exam findings:
- Stable vital signs.
- Dried blood on the lips.
- Hepatosplenomegaly.

Pertinent Laboratory findings:
- Platelet count 0 (baseline platelet count 35,000).
- Urine analysis - gross blood

Cytopathology:

Clinical Course:
- Day 1: Bleeding oral ulcers, Platelets 0, transfusion of 4 units platelets.
- Day 2: Diagnosis of Secondary Immune Thrombocytopenic Purpura (ITP) was made, started intravenous immunoglobulin (IVIG). No significant improvement.
- Day 4: Dexamethasone was started; discontinued on day 18 as no benefit.
- Day 6: Received Rituximab. Romiplostim infusions.
- Day 6: Developed Transfusion-related acute lung injury (TRALI) following platelet transfusion needing ICU transfer and mechanical ventilation.
- Day 8,10,14: Splenic artery embolized three times.
- In spite of all efforts platelet counts improved only transiently with worsening bleeding.
- Day 15: Laparoscopic splenectomy performed following which bleeding subsided and platelet counts improved to 100,000.
- Eventually discharged home and current platelet counts are within normal limits.

Discussion

This patient had chronic thrombocytopenia from cirrhosis and splenomegaly but subsequently developed secondary ITP from Hepatitis C.

Epidemiology: Incidence 100 per million

Figure 1: Peripheral blood smear showing complete lack of platelets, no schistocytes.

Clinical Course:

Figure 2: Platelet Count by Day

Pathogenesis of ITP:
Production of specific IgG autoantibodies by the patient’s B lymphocytes, most often directed against platelet membrane glycoproteins such as GPIIb/IIIa [3,4,5]

Diagnosis and treatment:
ITP is a diagnosis of exclusion[2,3]. Obtain good history regarding medical conditions like liver disease, thyroid disease; review medications; look for infectious causes especially viral such as HIV, HCV [6].

Figure 3: Estimated fraction of various forms of ITP.

Summary Points

1. Clinicians should expand the differential diagnosis of thrombocytopenia when it is acute or changing, especially to include diagnoses that are likely to harm the patient such as ITP, TTP (Thrombotic Thrombocytopenic Purpura) and DIC.

2. ITP is a diagnosis of exclusion.

3. Initial treatment of ITP includes steroids and IVIG.

4. In refractory ITP, splenectomy is very effective and is shown to cause sustained remission.

References


Figure 4: Treatment algorithm for ITP

Figure 5: Frequency of complete remission in ITP following Splenectomy

Conclusion

Splenectomy is the preferred therapy for patients with ITP who are refractory to first-line therapy with glucocorticoids or IVIG. Splenectomy leads to sustained remission in two-thirds of patients. Perioperative mortality rates for laparoscopic and open splenectomy are 0.2 and 1 percent, respectively [1].

Data for all 47 case series of 15 or more adults who had splenectomy for ITP, 1966-2004, with follow-up of 1 to 153 months (median, 24 months). The data demonstrate that the rate of complete remissions (defined as a normal platelet count with no further treatment for the duration of observation) did not change with time after splenectomy.

Figure 5: Frequency of complete remission in ITP following Splenectomy

Zero to Sixty in 48 Hours

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