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Immune Thrombocytopenia (ITP)

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Introduction:

Immune thrombocytopenia (ITP), formerly known as idiopathic thrombocytopenic purpura, is a haematological disorder characterized by a low platelet count in the blood, leading to an increased risk of bleeding and bruising. ITP is an autoimmune condition in which the body's immune system mistakenly targets and destroys its own platelets, essential blood components responsible for normal clot formation and hemostasis.

ITP presents a complex and multifaceted clinical landscape, challenging both patients and healthcare providers. This disorder can affect individuals of all ages, with distinct clinical manifestations and outcomes that vary widely among patients. While ITP primarily manifests as a bleeding disorder, it also has an emotional and psychological impact on those living with the condition, often leading to a diminished quality of life.

Pathophysiology

ITP is primarily a disease of increased peripheral platelet destruction mostly due to antibodies to specific platelet membrane glycoproteins. Relative marrow failure may contribute to this condition. Acute ITP often follows an acute infection with spontaneous resolution.

Etiology

ITP develops when platelets become coated with immunoglobulin G (IgG) autoantibodies to platelet membrane antigens resulting in splenic sequestration and phagocytosis by mononuclear macrophages resulting in a shortened life span of platelets in the circulation.

Incidence

Adults 66 per 1000000 / per year

Children 50 per 1000000/ per year

Chronic refractory ITP 10 per 1000,000 per year

Mortality

Hemorrhage most serious complication. ICH is the most significant.

Mortality from ICH. 1% in children, and 5% in adults.

Older age and previous history of haemorrhage increase the risk of severe bleeding.

Sex – Children – equal

Adults – female to male 2.6:1

Clinical presentation

Children's history of preceeding fever for a few weeks or live virus immunisation.

Adults:- Gradual

Purpura

Menorrhagia

Epistaxis

Gingival bleeding

Easy bruising

Evidence of ICH

D/D

1. Pseudo thrombocytopenia (Platelet Clumping)
2. Liver disease
3. Myelodysplasia
4. Lymphoproliferative diseases, autoimmune

5. Drug-induced (Alcohol, Heparin, Quinine, Sulfonamides)
6. Infection- HCV, HIV & B Virus / CMV / Corona
7. Malignancy
8. Megaloblastic anaemia
9. Transfusion-related

Lab

- CBC
- Peripheral smear
- Clotting time normal

Treatment

i. Platelet transfusion
ii. IV IG
iii. Steroids
iv. Antifibrinolytic agents – Tranexamic acid, Aminocaproic acid
v. Rituximab
vi. Splenectomy when medical therapy fails.
vii. Thrombopoietin receptor agonists (TPO-RA)

A 43-year-old female patient first reported to the hospital in May 2009 with complaints of bleeding ulcers over the tongue and rash over the arms and legs. She gave a history of body aches for 4-5 days and a fever of 1 day duration. Had been having a fever on and off for one month. No overt bleeding. Consulted a local doctor who advised blood tests which revealed very low platelet counts and advised a haematology consult.

She had a known case of hypothyroidism on replacement therapy for 15 years. Had 3 children by normal delivery. Attained menopause. Had pulmonary TB in 1994 treated and recovered.

Clinically mild fever, vitals stable. Petechiae present in the oral cavity, arms and legs. No organomegaly. Other systemic examination NAD. Investigations revealed Hb – 8.0Gram, TC 9000, platelets 20000. S.Bilirubin 2.4 others normal. USG abdomen – Fatty liver. Treated with antibiotics empirical antimalarials and support. Clinically improved but platelet count remained low.

Administered 2 doses of Methylprednisolone 500mg IV. Platelet count improved to 45000 and the rash subsided. The patient was asymptomatic and discharged on Eltroxin and vitamins advised review after 2 weeks. Readmitted 2 days after discharge with complaints of a rash over the tongue. No fever. No oral bleeding. Outside platelets 2000, TC 6000. Clinically stable. Platelets 3000. Started on IV Methylprednisolone 250mg IV OD x 5 days with good improvement. Discharged with a platelet count of 40000 on an oral tapering dose and was advised to review after 15 days with repeat investigations.

She was brought to the casualty after 20 days in an unconscious state. History obtained from relatives that the patient went to sleep, as usual, the previous night but did not wake up. Had been having headaches and vomiting for the past 2 days. No history of falls, history of giddiness and double vision since the previous day afternoon and unsteady gait since evening. No history of tonic-clonic seizures, rash or bleeding. Clinically stuporous state, 6th cranial nerve palsy, withdraws limbs to pain, plantar bilateral extensor. CT brain bilateral acute on chronic subdural haematoma. No mass effect. Platelet 49000. The patient started on Inj. Dexamethasone.

The Neurologist and haematologist advised to continue the same. Progressively

improved. Became alert, headache and diplopia improved. Clinically no deficit. Repeat CT after 8 days resolving bilateral chronic subdural hematoma. Platelets 1,69,000. Remained well. No neurological deficit. Discharged on a tapering dose of oral steroids and support. Reviewed once after discharge and was doing well. ANA profile showed SSA/RO52 positivity.

No further review till 2019 when she was admitted with UTI. Apparently had been continuing on low-dose steroids as advised and her platelets were normal.

Presenting a case of ITP who developed ICH treated symptomatically and had remained in remission after 6 months.

References

1. Tiede A, Sachs UJ, Czwalinna A, Werwitzke S, Bikker R, Krauss JK, et al. Prothrombotic immune thrombocytopenia after COVID-19 vaccination. Blood. 2021 Jul 29, 138 (4):350-353. [Medline]. [Full Text].
2. [Guideline] Neunert C, Lim W, Crowther M, Cohen A, Solberg L Jr, Crowther MA. The American Society of Hematology 2011 evidence-based practice guideline for immune thrombocytopenia. Blood. 2011 Apr 21. 117(16):4190-207. [Medline]. [Full Text].
3. Lambert MP, Gernsheimer TB. Clinical updates in adult immune thrombocytopenia. Blood. 2017 May 25. 129 (21):2829-2835. [Medline].
4. [Guideline] Neunert C, Terrell DR, Arnold DM, Buchanan G, Cines DB, Cooper N, et al. American Society of Hematology 2019 guidelines for immune thrombocytopenia. Blood Adv. 2019 Dec 10. 3 (23):3829-3866. [Medline]. [Full Text].
5. Provan D, Stasi R, Newland AC, Blanchette VS, Bolton-Maggs P, Bussel JB, et al. International consensus report on the investigation and management of primary immune thrombocytopenia. Blood. 2010 Jan 14. 115 (2):168-86. [Medline]. [Full Text].
6. Provan D, Newland AC. Current Management of Primary Immune Thrombocytopenia. Adv Ther. 2015 Oct. 32 (10):875-87. [Medline]. [Full Text].
7. Schultz CL, Mitra N, Schapira MM, Lambert MP. Influence of the American Society of Hematology guidelines on the management of newly diagnosed childhood immune thrombocytopenia. JAMA Pediatr.