AUTOIMMUNE COMPLEXITY: A CASE REPORT OF IMMUNE THROMBOCYTOPENIA (ITP) WITH COEXISTENT POSITIVE AUTOIMMUNE MARKERS AND MULTIDISCIPLINARY CHALLENGES

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ABSTRACT
This case study explores a complex clinical presentation of an 85-year-old male diagnosed with immune-mediated thrombocytopenia (ITP) with co-existent positive autoimmune markers. A multidisciplinary approach was crucial in managing the patient, involving hematology, rheumatology, urology, and respiratory specialists. Despite tailored treatments, including platelet transfusions, immunosuppressive therapies, and antibiotics, the patient developed complications including hospital-acquired pneumonia, pulmonary infection, and acute kidney injury. Extensive discussions with the family led to the decision to transition the patient to comfort care, emphasizing the challenges in managing complex cases and the importance of a comprehensive care plan.

Key Words: Immune Thrombocytopenia, Autoimmune Markers, Multidisciplinary Challenges, Elderly Patient, Haematological Disorders

INTRODUCTION

Immune-mediated thrombocytopenia (ITP) is the condition when the immune system mistakenly targets and destroys platelets in the body due to a recognition error (1). The exact cause of ITP remains unclear, but it is believed to involve the misrecognition of platelets by the patient's immune system, leading to their premature destruction (2). Recent population-based cohort studies indicate a heightened incidence of systemic lupus erythematosus among individuals with immune thrombocytopenic purpura (ITP) (3).

Case Scenario

An 85-year-old male patient with a history of the aforementioned medical conditions presented to Aga Khan University Hospital (AKUH) with a three-day history of per rectal, per oral bleeding, and mild bleeding from bed sores. Upon admission, a comprehensive clinical assessment and laboratory workup showed a low platelet count. This prompted a provisional diagnosis of immune-mediated thrombocytopenia.

In response to the ITP diagnosis, the patient received a tailored treatment regimen, which included platelet transfusions, intravenous fluids, IV tranexamic acid, and topical epinephrine to manage and control the bleeding episodes. Hematology specialists were actively involved in the patient's care, and further investigations led to a pivotal step in the diagnostic process.

A bone marrow biopsy was performed under local anesthesia. The final biopsy report suggested normocellular bone marrow with peripheral platelet destruction, indicating a potential immune-mediated origin of thrombocytopenia. In the light of this finding, the patient underwent a series of treatments, including three doses of pulse IV Methylprednisolone, which were subsequently transitioned to hydrocortisone as per hematology recommendations. Eltrombopag, a thrombopoietin receptor agonist, was also initiated to stimulate platelet production. A summary is given in Table 1 and 2.

Given the complex clinical presentation and the presence of positive antinuclear antibodies (ANA) and anti-double-stranded DNA (anti-ds-DNA), rheumatology specialists were consulted to explore

potential autoimmune associations (Table 3). Although the patient developed hematuria during hospitalization, the urology team advised against immediate intervention.

During the patient's stay, additional complications emerged, including hospital-acquired pneumonia, necessitating the optimization of antibiotic therapy. Expressly, tracheal cultures indicated a heavy growth of Klebsiella and Acinetobacter, leading to the initiation of IV Piperacillin-Tazobactam (Pip/taz). Furthermore, urine cultures identified E. coli, prompting adjustments to the antibiotic regimen to include IV Meropenem, IV Vancomycin, and IV Colistin. The patient's respiratory status deteriorated, leading to increased oxygen requirements and persistent desaturation. As a result, the patient was transitioned to Bilevel Positive Airway Pressure (BIPAP) therapy and transferred to the Special Care Unit for closer monitoring. A repeat chest X-ray revealed a worsening of bilateral perihilar patchy airspace opacities, indicative of a superimposed pulmonary infection.

Final blood culture reports identified Stenotrophomonas Maltophilia, necessitating further optimization of antibiotic therapy. The patient received multiple platelet transfusions throughout the hospital course and underwent three packed red blood cell (PCV) transfusions. Notably, the patient experienced acute kidney injury (AKI), which responded positively to gentle hydration.

Amidst the evolving medical challenges, detailed discussions were made with the patient's family regarding prognosis and treatment options. Ultimately, the family decided to transition the patient to comfort care. Considering the ongoing drop in platelet counts, rheumatology specialists discussed the potential use of intravenous immunoglobulin (IVIG) with the family, ensuring comprehensive communication and decision-making.

Following extensive dialogue with the patient's family, it was decided to discharge the patient with a structured nursing care plan. This included oral prednisolone, eltrombopag, and weekly complete blood counts (CBC) monitoring. The family was also advised to seek immediate medical attention at AKUH-ER in the event of any bleeding episodes or emergent medical concerns.

Associated Diagnosis/Significant Comorbid

The patient under consideration exhibits a medically intricate profile featuring Chronic Liver Disease (CLD) of non-B and non-C origin, a known history of hypertension, a previous diagnosis of benign prostatic hyperplasia (BPH) successfully managed with transurethral resection of the prostate (TURP), a prior non-ST-segment elevation myocardial infarction (NSTEMI) with a preserved ejection fraction (EF) of 55% under medical management, a documented abdominal aortic aneurysm, concurrent peripheral artery disease (PAD), and a pre-existing motor neuron disease.

Discussion

This patient's presentation of ITP, characterized by a low platelet count and positive autoimmune markers (ANA and anti-ds-DNA), is intriguing. ITP is known for its autoimmune pathophysiology, where the immune system erroneously targets and destroys platelets. The co-occurrence of ITP with positive autoimmune markers raises questions about potential associations with systemic autoimmune disorders. Recent studies have shed light on the management of complex cases of ITP. In a study in 2020, the authors explored the clinical characteristics and outcomes of ITP associated with COVID-19, emphasizing the importance of a multidisciplinary approach in managing patients with both autoimmune conditions and infectious diseases (5). Furthermore, it was updated with international consensus guidelines for investigating and managing primary ITP, providing valuable insights into the diagnosis and Treatment of this condition (4). The positive ANA and elevated dsDNA levels suggest potential autoimmune involvement beyond ITP (3).

Table 1. Pharmacological Treatment of the patient after Discharge (Take Home Medicines)

Drug	Dose	Route	Frequency	Duration of treatment
Tab Calcium with Vitamin D	1 Tablet	Chewa-ble	Once a day (4)	Continue till next follow-up
Bag Ceftazidime 1000 mg/bag	2000 mg	IVPB piggy-back	Every 12 hours	Continue till 07/09/2023
vl Ipratropium bromide 500 mcg/2ml	500 mcg	Nebu-lizer	Every 08 hours	Continue till next follow-up
Tab Eltrombopag 50 mg/tab	100 mg	Oral	OD	Continue till the next follow-up
Cap Itopride Hydrochloride 150 mg/cap	150 mg	Oral	OD	Continue till next follow-up
Tab Atorvastatin Calcium 10 mg/tab	10 mg	Oral	At bed time	Continue till next follow-up
Tab Metoclopramide HCL 10 mg/tab	10 mg	Oral	Before meals	Continue till next follow-up
Sch Polyethylene glycol (peg) 1 sac	1 sac	Oral	OD	Continue till next follow-up
Tab Prednisolone 5 mg/tab	15 mg	Oral	Two times a day	Continue till next follow-up
Cap Omeprazole 20 mg/cap	40 mg	Oral	Before breakfast	Continue till the next follow-up

Table 2.0. Laboratory Investigations of the Patients in the Internal Medicine Department

Date	20/8/23	21/8/23	22/8/23	23/8/23	24/8/23	25/8/23	26/8/23	27/8/23	28/8/23
Hb	8.9	8.9	9.4	8.6	10.0	9.1	9.6	9.1	9.1
HCT	28.1	28.2	29.5	28.0	31.4	29.1	29.9	30.2	30.2
MCV	91.2	92.2	92.8	93.3	92.1	93.6	93.2	93.8	98.1
WBC	26.1	27.8	27.3	30.5	31.9	26.8	23.0	26.6	27.3
Neu	93.4	96.1	95.2	96.3	94.3	92.9	95.5	95.9	95.6
Lym	1.1	0.9	1.1	1.3	1.3	2.2	1.2	1.1	0.9
Platelets	14	09	43	26	49	38	17	22	19
BUN	47	47	53	60		78		74	74
Creatinine	1.4	1.2	1.2	1.4		1.8	1.9	1.7	1.5
Na		142	143	145		148	150	153	146
K	4.3	4.4	4.8	4.5	4.2	4.3	4.1	5.0	4.6
Cl		107	106	106	109	109	113	114	109
BIC		26.2	26.5	26.1		27.4	26.2	28.9	24.9
Ca						8.3	8.3		
Mg						3.0			
Alb						3.3			
CRP							17.2		12.3

Table 3. Autoimmune profile of the patient

Lab Investigation	Value
Coomb's Test	2+
ANA	Positive
dsDNA	37.6

Conclusion

This case underscores the complexities inherent in managing elderly patients with immune thrombocytopenia and concurrent autoimmune markers, especially in the presence of multiple comorbidities. The coexistence of ITP with positive antinuclear antibodies (ANA) and anti-double-stranded DNA (anti-ds-DNA) raises questions about potential associations with systemic autoimmune disorders. The multidisciplinary approach proved pivotal in addressing evolving medical challenges, but despite interventions, the patient's deteriorating health necessitated a transition to comfort care. This emphasizes the need for ongoing research and guidelines to navigate the intricate landscape of autoimmune complexities in the elderly population, ensuring comprehensive and personalized patient care.

References

1. Donoghue E. Immune-Mediated Thrombocytopenia 2019.
2. Zufferey A, Kapur R, Semple JW. Pathogenesis and Therapeutic Mechanisms in Immune Thrombocytopenia (ITP). J Clin Med. 2017;6(2).
3. Pamuk ON, Ali SM, Hasni S. Development of systemic lupus erythematosus in patients with immune thrombocytopenic purpura: A systematic meta-analysis. Autoimmun Rev. 2023;22(4):103297.
4. Provan D, Arnold DM, Bussel JB, Chong BH, Cooper N, Gernsheimer T, et al. Updated international consensus report on the investigation and management of primary immune thrombocytopenia. Blood advances. 2019;3(22):3780-817.
5. Mahévas M, Moulis G, Andres E, Riviere E, Garzaro M, Crickx E, et al. Clinical characteristics, management and outcome of COVID-19-associated immune thrombocytopenia: a French multicentre series. British journal of haematology. 2020;190(4):e224-e9.