Original Research Paper

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General Medicine

ADULT EVAN'S SYNDROME CASE

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KEYWORDS :

INTRODUCTION

Evans syndrome is an autoimmune condition that presents with 2 or more cytopenia's which commonly includes autoimmune haemolytic anaemia and immune thrombocytopenia, with or without neutropenia . Classification – Primary (idiopathic) Secondary – a/w SLE, autoimmune lymphoproliferative syndrome, (ALPS),common variable immunodeficiency (CVID), CLL, NHL (patients age>50 yrs, viral infections (HIV, hepatitis C) , following allogenic hematopoietic stem cell transplant.

Case History

A 51 yrs old female which was presented in emergency department at MGM hospital, Chhatrapati Sambhaji Nagar, with viral fever and acute respiratory distress syndrome, severe anaemia and jaundice. She had h/o multiple packed cell volume and Random donor platelets transfusion. She has history of thrombocytopenia since 6 years, she is also a known case of hypothyroidism on medical treatment.

Chief complaints :-
Breathlessness since 3 days
Chest pain since 3 days
1 episode of hematemesis 3 days back

History Of Present Illness :-

Patient was apparently alright 3 days back then had sudden onset of breathlessness which was progressive in nature associated with chest pain. She also had an episode of hematemesis.

Patient was admitted outside hospital for general weakness, where routine labs were done along with upper gastrointestinal endoscopy and bone marrow biopsy was done.

She also had Malena 7 days ago.

No h/o petechiae

No h/o ecchymosis

▪ **Past h/o –** Hypothyroidism since 7 years and on medical treatment.

h/o of multiple packed cell volume and random donor platelet transfusion

General Examination

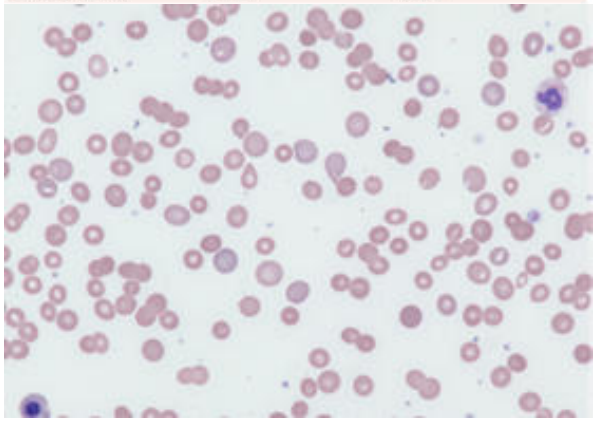
Height- 156 cm
Weight- 50kg
BMI- 20.5
Temperature- Afebrile
Pulse- 109 bpm, regular
Pulse pressure- 60 mmHg
Peripheral pulsations -felt
Blood pressure – 140/80 mmHg
Pallor- Present
Icterus- Mild
Clubbing- Absent
JVP- raised
Edema- Absent
Lymphadenopathy- Absent

Systemic Examination

1) Respiratory system- Breath sounds symmetrical
Bilateral crepitations heard
2) Abdominal system- Mild splenomegaly
No hepatomegaly
3) Cardiovascular system – Normal (S1 +, S2+ heard)
4) Central Nervous System – Normal

Parameter	Normal values	Day 1	Day 2	Day 4	Day 6	Day 7	Day 8	Day 9	Day 11	Day 12
Hb	12-16.2 gm/dl	9.2	7.7	7.6	7.8	6.8	8	8.4	8.8	8.7
TLC	3500-9060/cumm	21950	14,700	14,000	22,000	18,800	18,600	22,100	17,800	16,300
PLC	1,65,000-4,15,000/cumm	5,000	7,000	6,000	9,000	12,000	18,000	44,000	1,10,000	1,46,000
Urea	7-20 mg/dl	60	-	96	-	80	51	-	27	-
Serum Bilirubin	0.3-1.3 mg/dl	4.10		4.4						
Direct Bilirubin	0.1-0.4 mg/dl	0.7		0.6						
Indirect Bilirubin	0.2- 0.9 mg/dl	3.4		3.8						
Serum Creatinine	0.5-1.2 mg/dl	0.6		0.8		0.7	0.5		0.5	

Parameters	Normal range	Observed value
Serum Iron	50-170 ug/dl	**189**
Total iron binding capacity	250-450 ug/dl	**248**
Transferrin saturation	20-50%	**76.21**
LDH	120-246 U/L	**1086**
Free T3	2.3-4.2 pg	2.15
Free T4	0.8- 2.19 ng/dl	1.14
TSH	0.4-4.6 mIU/ml	1.48
Reticulocyte count	0.2-2%	**8.60 %**



Peripheral smear showed **spherocytes** with marked **anisopoikilocytosis**

Management Of Patient

Aspect to be managed	Management
Severe anaemia	Packed cell volume transfusion initially outside hospital
Severe thrombocytopenia (< 10,000/cumm)	Random donor platelet
Autoimmune disorder	Methyl Prednisolone pulse therapy 1 g /day for 3 days then shifted oral 1mg/kg for 1 week then tapered gradually
Viral fever (H1N1 / any other virus causing LRTI)	Anti-virals (Oseltamivir i/v)

Special Test Performed
1) Direct Coombs Test- Positive (++)
2) Bone marrow biopsy - Normal cellularity
3) Anti Nuclear Antibody - ++ (Moderate positive)

How We Reached The Diagnosis
1) Confirm AIHA (Direct coombs test positive, positive antinuclear antibodies, raised LDH, Peripheral smear)
2) Thrombocytopenia
3) Exclude secondary causes- lymphoproliferative disease, infections, primary immunodeficiency (CVID) , Antiphospholipid antibody syndrome

Diagnosis-
AIHA and ITP without an identifiable secondary cause strongly suggests Evan's Syndrome.

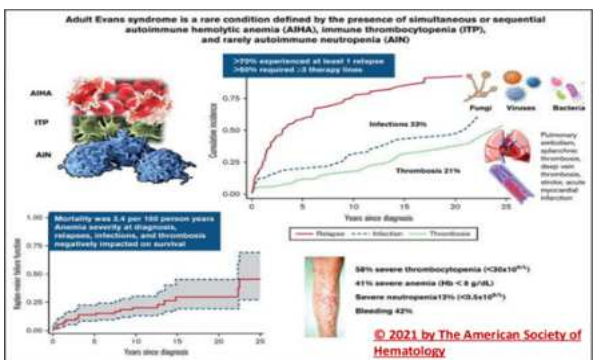
Outcome And Follow Up
Patient was discharged when Hb and serum Bilirubin were in normal range and Platelet count was more than 1,00,000/cumm

Patient is now stable and responding well to steroids with improvement in anaemia and thrombocytopenia.

Aetiology
Idiopathic
Excessive immune dysregulation
B cells → auto Ab → against platelet , RBC , WBC

Signs and symptoms of patients can be variable depending on type of blood cell line affected
1) AIHA – fatigue, pallor, dizziness , breathlessness, jaundice, splenomegaly
2) ITP- Easy bruising, petechiae, purpura
Increased risk of – Ischemic complications (ACS, CVA secondary to AIHA in patients > 60 yrs,
life threatening haemorrhage)

Epidemiology
Rare condition
Diagnosed in less than 5% of all patients with either AIHA , ITP at the onset
Mean age at the time of diagnosis – 52 years
F >M
3:2
Children > adults
No genes associated
Rarely can be inherited - Familial Evans syndrome
Co-occurs with Heart defects or inherited along with hereditary spastic paraplegia



Investigations
A] Evaluation of hemolysis-
1) LDH
2) Haptoglobin
3) Bilirubin
4) Retic count
B] PS –
1) Direct coombs test
2)Spherocytes
(confirms warm AIHA)
PS rule out cold agglutinin ,TTP
C] Ix for infectious causes (HIV, Hep C), other autoimmune diseases(SLE, ALPS, etc.)and malignancies (CT ,CXR) should be done before diagnosis of Evans syndrome can be made.

1st line Corticosteroids (PREDNISOLONE 1-2mg/kg) IVIG (intravenous immunoglobulins)	→ For ITP (tapered over weeks for ITP / months for AIHA) → IVIG is more preferred for AIHA
2nd line RITUXIMAB (CD 20 inhibitor) DANAZOL	→When Evans syndrome is secondary to autoimmune / malignancy → Used when there are corticosteroid sparing effects
3rd line (tried drugs – immunosuppressants) CYCLOPHOSPHAMIDE MYCOPHENOLATE MOTEFIL AZATHIOPRINE CYCLOSPORINE SIROLIMUS	Dependant on patient factors , co-morbidities and autoimmune conditions)
Last line Hematopoietic stem cell transplant (autologous /allogenic)	Last option in those unresponsive to all medical treatments

CONCLUSION
Adult ES is often severe and marked by a relapsing clinical course and potentially fatal complications, pinpointing the need for high clinical awareness, prompt therapy, and anti-infectious/anti-thrombotic prophylaxis.

REFERENCES
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2) © 2021 by The American Society of Hematology
3) Harrison's principle of internal medicine