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(CASE REPORT)

Unusual presentation of Evans syndrome with hypokalemic paralysis during pregnancy, management and outcome

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Abstract

Background: Evans Syndrome (ES) is a rare autoimmune disorder characterized by autoimmune hemolytic anemia (AIHA) and immune thrombocytopenic purpura (ITP). Its occurrence in association with Sjögren's Syndrome (SS) during pregnancy, compounded by hypokalemic paralysis, is exceedingly rare.

Case Description: A 24-year-old primigravida at 17 weeks of gestation presented with limb weakness due to hypokalemic paralysis. She had a history of ES and SS diagnosed prior to conception, managed initially with corticosteroids and vitamin B12 supplementation. During pregnancy, she experienced hypokalemia, renal tubular acidosis, and moderate anemia, which were managed effectively with potassium, bicarbonate, prednisolone, and hydroxychloroquine. The pregnancy was monitored closely, culminating in a cesarean delivery of a healthy baby at 39 weeks. Postpartum care included transfusions and anticoagulation therapy. Both mother and baby showed favorable outcomes without major complications.

Conclusion: The coexistence of ES and SS during pregnancy, further complicated by hypokalemic paralysis, presents unique challenges. Early diagnosis, multidisciplinary management, and close monitoring are pivotal in achieving successful maternal and neonatal outcomes.

Keywords: Evans Syndrome; Sjögren's Syndrome; Pregnancy Complications; Hypokalemic Paralysis; Autoimmune Hemolytic Anemia (AIHA)

1. Introduction

Evans syndrome (ES) is a rare autoimmune disease characterized by autoimmune hemolytic anemia (AIHA) and immune thrombocytopenic purpura (ITP)[1]. ES can be primary or secondary to another autoimmune disease most commonly systemic lupus erythematosus (SLE), rarely with primary Sjogren's syndrome (SS). Hypokalemic paralysis manifested as acute muscular numbness/ weakness to paralysis during pregnancy is an extremely rare occurrence.

2. Case report

A 24 yr old primigravida with 17 weeks live pregnancy complained of weakness of both upper and lower limbs and bedridden, vitals were stable, Power 4/5 in all extremities, bilateral flexor plantar, sensation were normal bilaterally, hypoactive deep tendon reflexes.

She was diagnosed 3 months before conception with Sjogren's syndrome with a positive antinuclear antibody (ANA 1:90, nuclear fine speckled 3+, cytoplasmic granular 3) anti-SSA, anti-Ro 52 (fig-5,6) and Evans syndrome: warm AIHA

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with severe anemia in heart failure with vit B12 deficiency(fig-3), (fig-4) and managed with pulse Dexamethasone 40mg for 4 days, 1 pint PRBC transfused, 4 days of Vit B12 injections following which patients blood counts improved (fig-1) She was discharged on Prednisone with calcium, multivitamin and folic acid. She stopped taking steroids from end of first trimester without follow up. At present her lab investigation showed (fig-3)low potassium levels , renal tubular acidosis associated low bicarbonate levels ,hypocortisolemia, no features of hemolysis, LDH, indirect bilirubin was in normal range, with moderate anemia ,peripheral smear showed macrocytic blood picture.

She was started on potassium, magnesium, bicarbonate, prednisolone and Hydroxychloroquine with Inj vitamin B12 and oral iron supplements. Patient's weakness improved.

Patient was on remission on prednisolone and Hydroxychloroquine which was continued throughout pregnanc. Patient came for regular antenatal visits and were uneventful.

An emergency LSCS for cephalopelvic disproportion in labour at 39 weeks of gestation was done, delivered an healthy baby of weight 3.3 kg with bilateral club foot.

Postoperatively for Hb 7.5g/dl(fig-2)1 prbc transfused ,antibiotic, tab prednisolone, hcq, syp potassium, Inj enoxaparin 20mg/0.2ml and tab aspirin 75mg od was started and discharged with same drugs for 6 weeks.

Patient is on follow up and on remission. Baby has normal blood parameters.

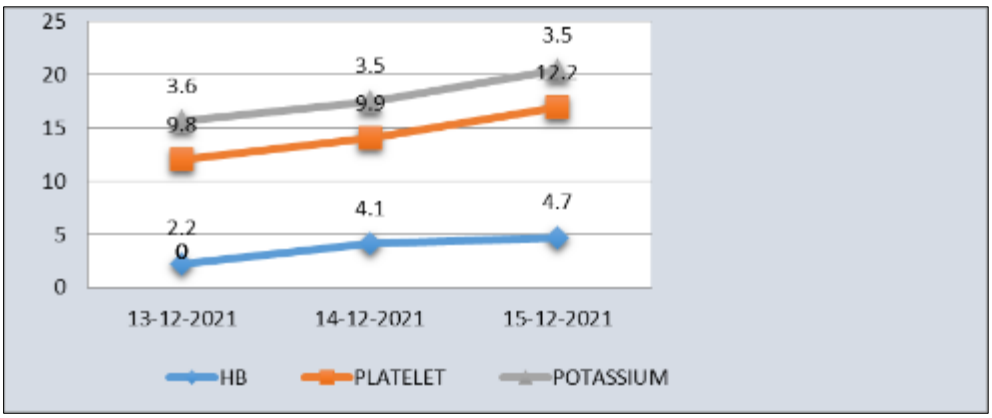

Figure 1 Trends in Hemoglobin, Platelet Count, and Potassium Levels Over Time

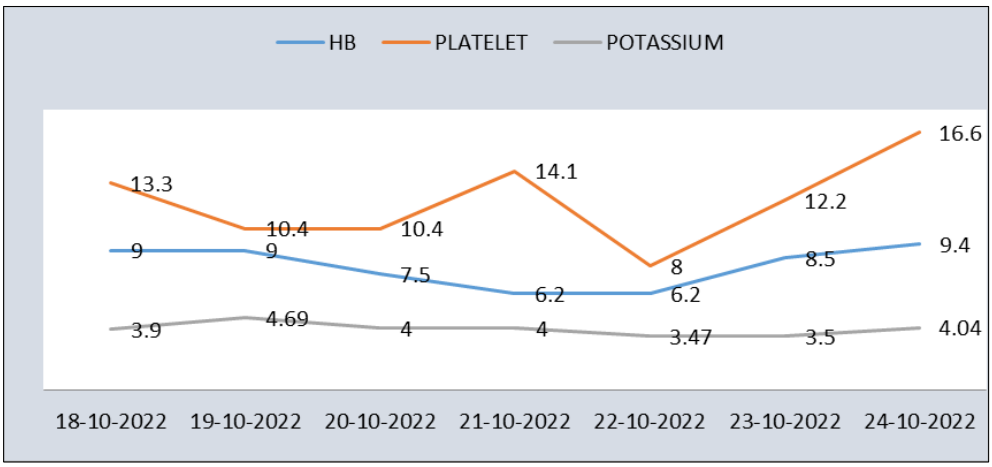

Figure 2 Fluctuations in Hemoglobin, Platelet Count, and Potassium Levels Over a Week

	13-12-2021	14-12-2021	15-12-2021	13-05-2022	14-05-2022	15-05-2022
HB	2.2	4.1	4.7	7		
PLATELET	98	99	122	1.44	1.46	1.36
S.POTASSIUM	3.6			2.93	4.1	3.16
T.BILIRUBIN	2.09			0.38		
D.BILIRUBIN	0.89			0.18		
INDIRECT BILIRUBIN	1.2			0.2		
S.LDH	599					
S.MAGNESIUM				2.1		
BICARBONATE				11	11.6	17.8
B12	10.8					

Figure 3 Biochemical and Hematological Parameters Recorded Over Time

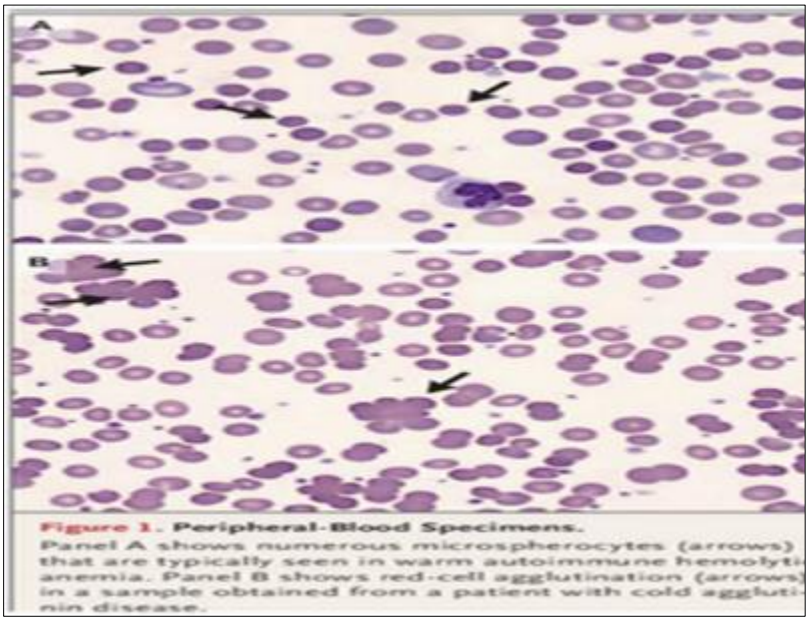

Figure 4 Peripheral blood smear

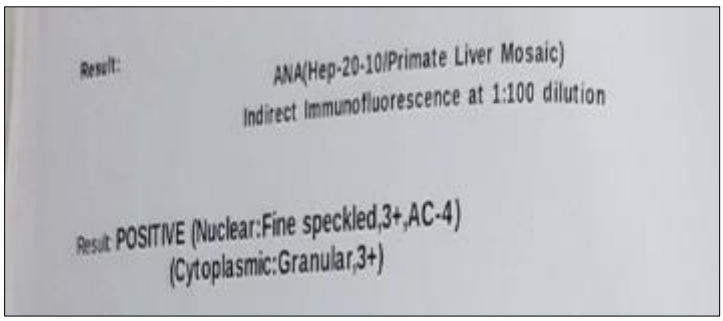

Figure 5 ANA report positive

Antigen	Intensity	Class
RNP/Sm	1	0
Sm	1	0
SS-A native (80 kDa)	98	.+++
Ro-52(52 kDa)	111	.+++
SS-B	5	0
Scl-70	0	0
PM-Scl	0	0
Jo-1	3	0
Centromere B	0	0
PCNA	2	0
dsDNA	0	0
Nucleosomes	0	0
Histones	3	0
Ribosomal-P Protein	2	0
AMA-M2	0	0
Control	97	.+++

Figure 6 Autoantibodies profile

3. Discussion

Evans syndrome is a rare autoimmune disorder characterized by simultaneous or sequential presence of a positive antiglobulin test, autoimmune haemolytic anemia (AIHA), and immune thrombocytopenia (ITP). It is characterised by frequent exacerbations and remissions within a chronic course. Evans syndrome association with Sjogrens Syndrome is rare. Similiarly we had ES in 0.02 % (1 /4800 live births).

Pregnancy in association with AIHA may provoke life-threatening anaemia in 40-50% of cases, and stillbirth or severe postpartum haemolytic anaemia in 35- 40% of infants[2], in our case we had life threatening anemia, to worsen the condition we encountered hypokalemic paralysis which has so far not been reported in literature.

In study conducted by Manjula SK, 1 out of 4 patients had pre eclampsia, and none had abruption, PPH. There was one IUD, cause of which is unidentified[3], but in the present case we had none of the above complications.

In study conducted by Lefkou E ,2 out of 11 had still births, one due to intra cranial subdural hematoma, second one due to erythroblastic feature with evidence of auto immune hemolysis[4], however in our case we did not encounter any neonatal complications.

Hypokalemic paralysis with Evans and Sjogrens Syndrome, a case scenario never published, was successfully managed and pregnancy terminated at 39 weeks with a healthy baby.

4. Conclusion

Association of Evans syndrome with Sjogren's syndrome in pregnancy is a very rare disorder and its complication like hypokalemic paralysis should be kept in mind for differential diagnosis in patients presenting with unexplained thrombocytopenia during pregnancy. Close follow up, early management, careful planning and preparation for delivery in such women would enhance the chances of a favorable outcome.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

Ethical approval for this case report was obtained from the relevant institutional ethics committee, ensuring adherence to ethical guidelines for research involving human participants.

Statement of informed consent

Written informed consent was obtained from the patient for the publication of this case report, including all clinical details and any accompanying images, with assurance of anonymity and confidentiality.

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